CHAPTER 11

Intraocular Tumours

Iris tumours

Iris melanoma

In general, uveal melanomas are three times commoner in patients with blue/grey than brown irides. They are extremely rare in black people and there is no sexual predilection. Conditions associated with or predisposed to early-onset uveal melanomas are: (a) *ocular melanocytosis*, (b) *naevus of Ota*, (c) *dysplastic cutaneous naevi*, (d) *familial melanoma* and (e) *neurofibromatosis-1*. Iris melanoma makes up about 5% of uveal melanomas. The majority are composed of spindle B cells (*see* below) and are of low malignancy. A minority contain an epithelioid cell component and can be aggressive. The tumour usually grows very slowly along the iris surface and may invade the angle and anterior ciliary body. The prognosis is very good and only about 5% of patients develop metastases.

Clinical features

1. **Presentation** is in the fifth to sixth decades, a decade earlier than ciliary body and choroidal melanoma, with enlargement of a pre-existing iris lesion.
2. **Signs**
   a. Typical
   - A pigmented or non-pigmented nodule at least 3 mm in diameter and 1 mm thickness located in the *inferior half* of the iris with a smooth or irregular surface (Fig. 11.1). Surface vascularity is also present and is easier to detect in a non-pigmented (Fig. 11.2) than a highly pigmented tumour, where it may be masked.
   - Pupillary distortion, ectropion uveae and occasionally localized cataract may be seen (Fig. 11.3).
   - Features associated with a prominent epithelioid cell component include prominent vascularity, rapid growth and heterogeneous pigmentation.



Fig. 11.2
Non-pigmented iris melanoma with prominent surface vessels

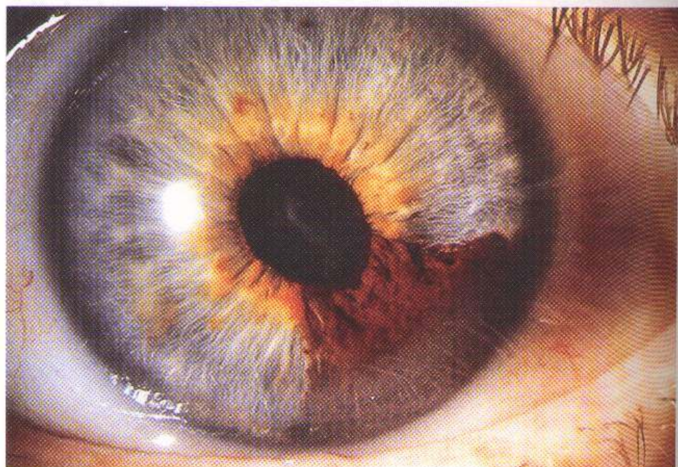


Fig. 11.3
Iris melanoma causing distortion of the pupil, ectropion uveae and a localized lens opacity

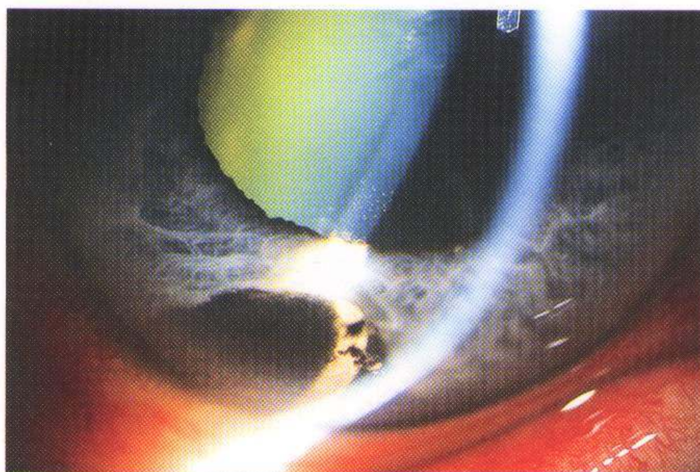


Fig. 11.1
Pigmented iris melanoma

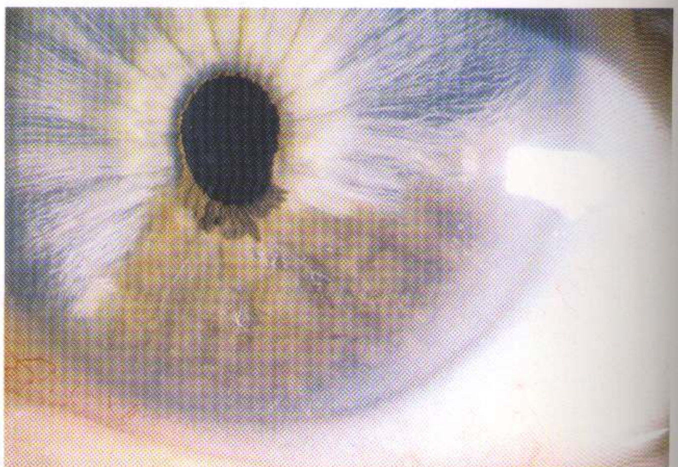


Fig. 11.4
Tapioca iris melanoma (Courtesy of B. Damato)

b. Rare variants

- Diffusely growing intrastromal melanoma may give rise to ipsilateral hyperchromic heterochromia.
- 'Tapioca melanoma' is characterized by multiple surface nodules (Fig. 11.4).

Treatment

1. **Observation** of suspicious lesions involves documentation by slit-lamp examination, gonioscopy and photography. Follow-up should be lifelong because growth may occur after several years of apparent inactivity. Initially the patient is reviewed after 3–6 months, then 6–9 months and later annually.
2. **Iridectomy** for small tumours with iris reconstruction to reduce postoperative photophobia.
3. **Iridocyclectomy** for tumours invading the angle (Fig. 11.5).
4. **Radiotherapy** with local plaques (brachytherapy) or external irradiation with charged particles for non-resectable tumours.
5. **Enucleation** may be required for diffusely growing tumours.

Differential diagnosis

1. **Iris naevus**, if large and distorting the pupil (*see* Fig. 11.11).
2. **Ciliary body melanoma** with extension through the iris root (*see* Fig. 11.24).
3. **Metastasis** to the iris is rare and usually occurs in patients with a known systemic malignancy. It is a pink or yellow, fast-growing mass (Fig. 11.6) which may be associated with anterior uveitis and occasionally hyphaema. Small, multiple deposits may also be seen (Fig 11.7).
4. **Adenoma of the iris pigment epithelium** is a rare benign tumour characterized by a dark grey-black nodule with a smooth but sometimes multinodular surface, most frequently in the peripheral iris (Fig. 11.8). The lesion causes anterior displacement and thinning of the iris stroma, which eventually erodes, disclosing the tumour on slit-lamp biomicroscopy.
5. **Leiomyoma** is an extremely rare benign tumour which arises from smooth muscle. The appearance is similar to that of an amelanotic melanoma except that it is not necessarily confined to the inferior half of the iris (Fig. 11.9). Often the diagnosis can be established only histologically.
6. **Primary iris cyst** (*see* below).

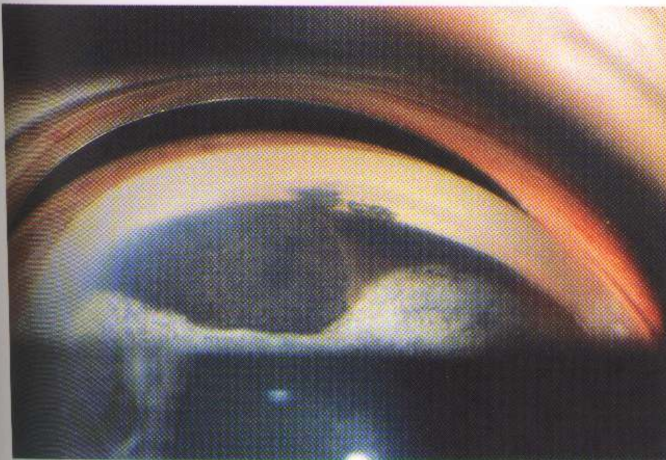


Fig. 11.5
Gonioscopic view of iris melanoma involving the angle

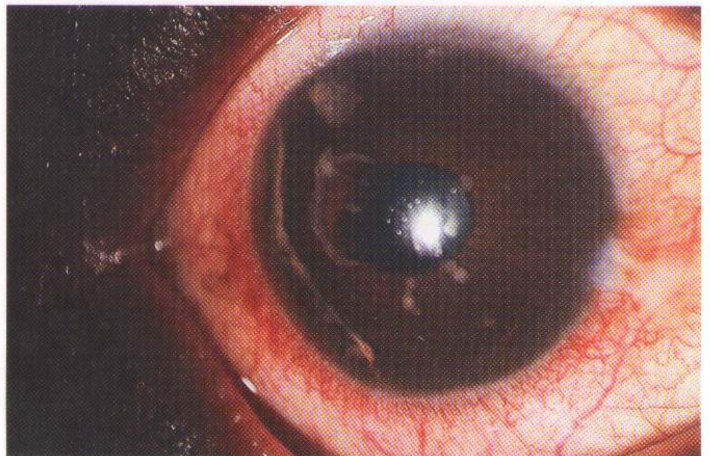


Fig. 11.7
Multiple iris metastases (Courtesy of B. Damato)

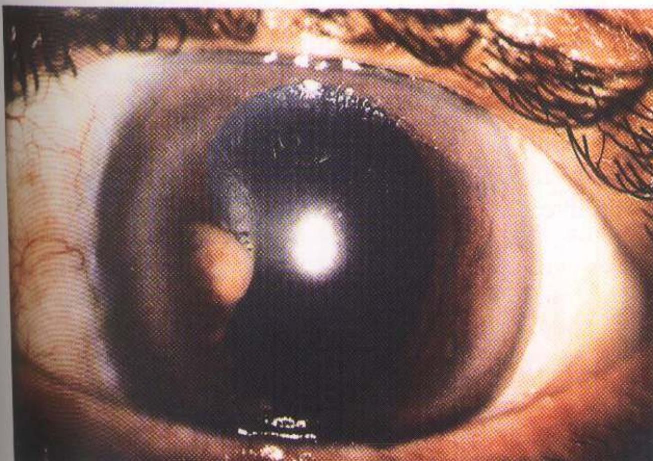


Fig. 11.6
Solitary iris metastasis

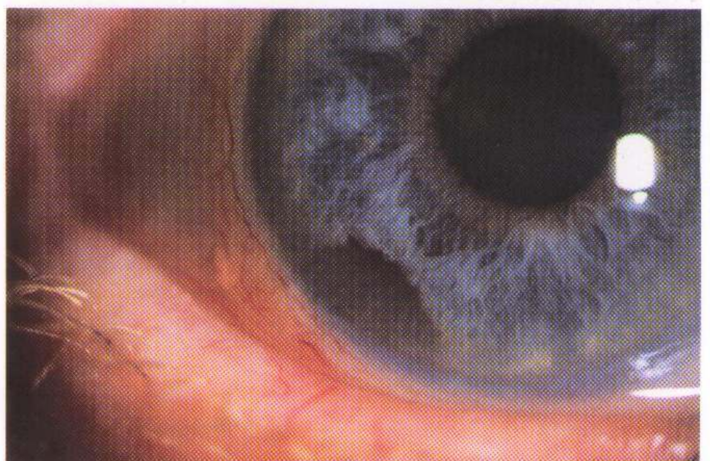


Fig. 11.8
Adenoma of the iris pigment epithelium

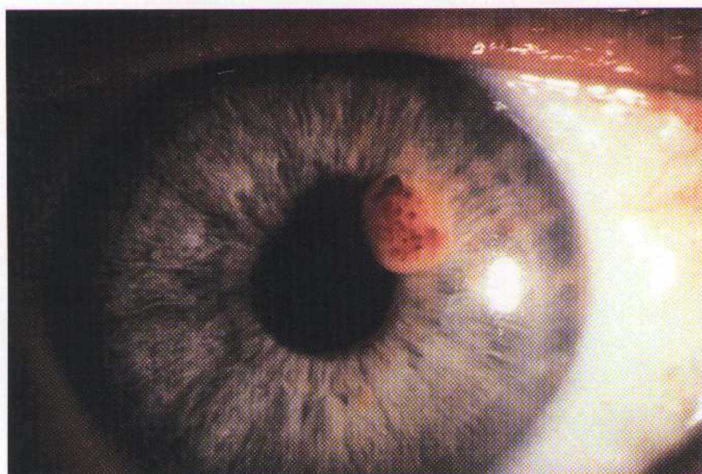


Fig. 11.9
Iris leiomyoma

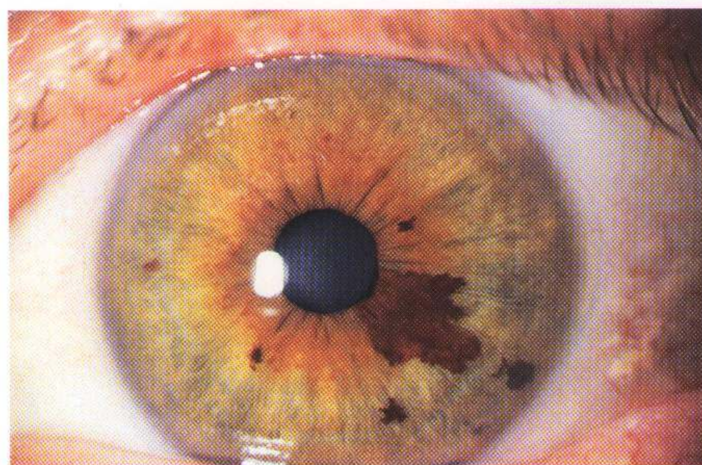


Fig. 11.10
Iris naevus and freckles



Fig. 11.11
Iris naevus distorting the pupil

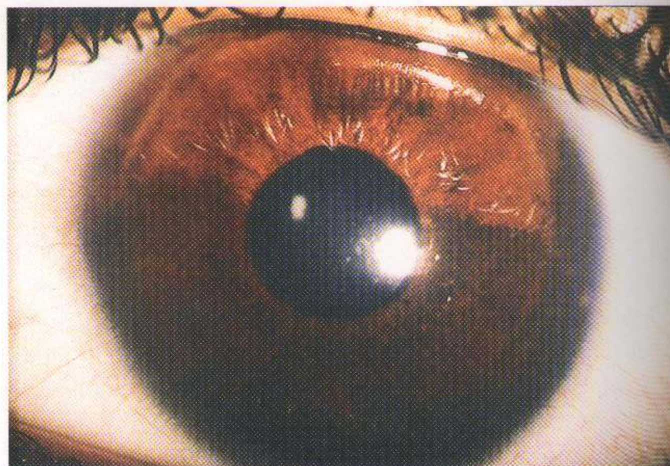


Fig. 11.12
Diffuse iris naevus

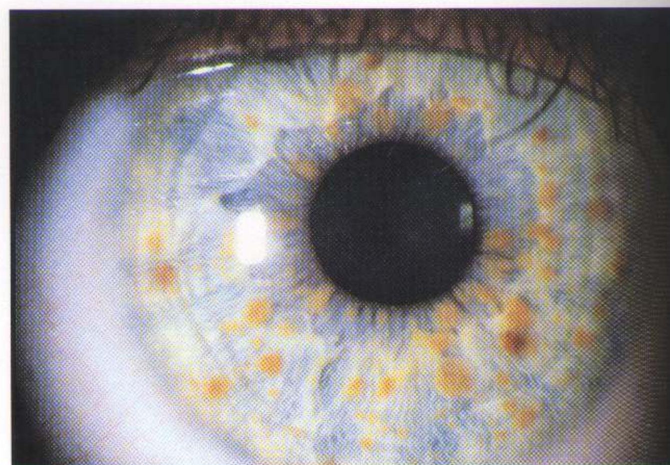


Fig. 11.13
Lisch nodules

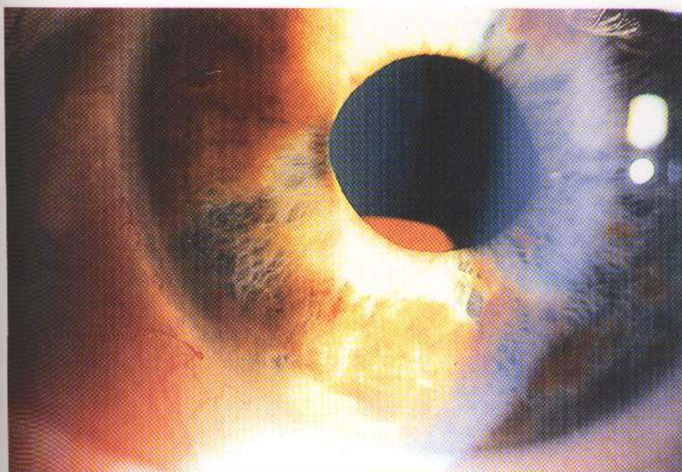
Iris naevi

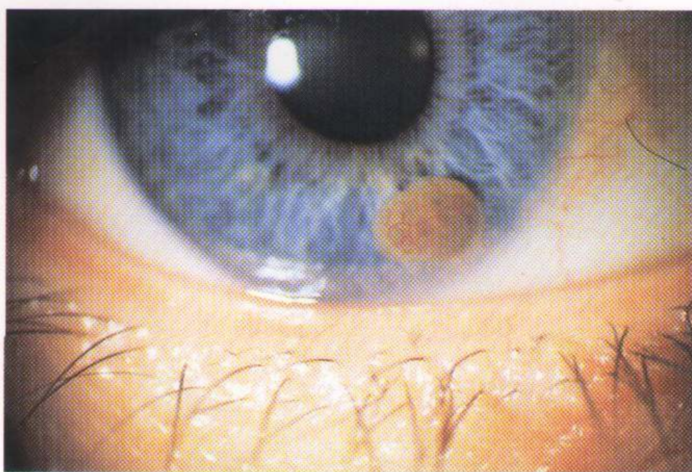
1. **A typical naevus** is a pigmented, flat or slightly elevated lesion, usually less than 3mm in diameter (Fig. 11.10). It is located in the superficial layers and may occasionally cause mild distortion of the pupil and ectropion uveae (Fig. 11.11).
2. **A diffuse naevus** obscures normal iris crypts and gives rise to hyperchromic heterochromia (Fig. 11.12). It may be seen in patients with the *Cogan–Reese syndrome* (*see* Chapter 9) and may also show small pedunculated nodules resembling mammillations.
3. **Lisch nodules** are multiple, small, bilateral, melanocytic hamartomas found after the age of 16 years in virtually all patients with neurofibromatosis-1 (Fig. 11.13).
4. **Freckles** are smaller than naevi (*see* Fig. 11.10). They are frequently multiple and bilateral, but never distort the iris architecture.

Iris cysts

Primary cysts

Primary iris cysts are rare curiosities arising from the pigment epithelium or, rarely, the stroma. The vast majority,


Fig. 11.14
Pupillary epithelial iris cyst


Fig. 11.16
Primary stromal iris cyst

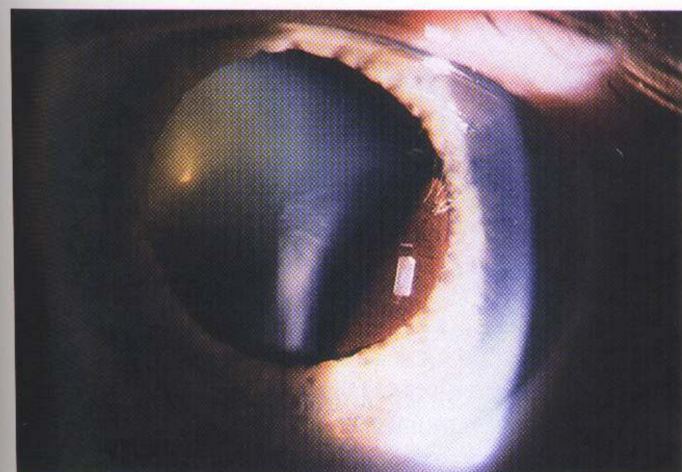
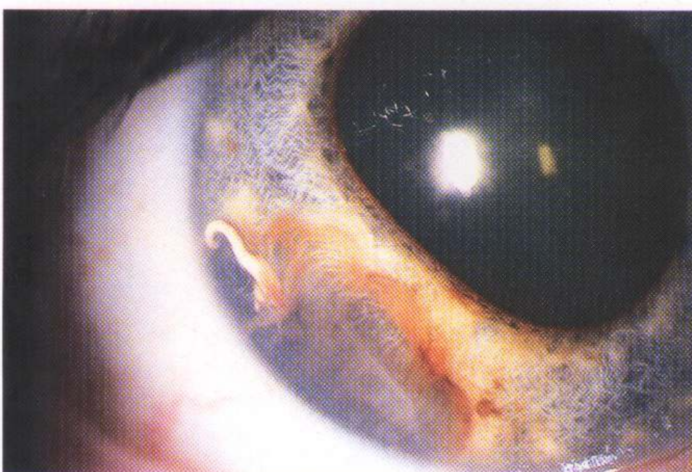

Fig. 11.15
Mid-zonal epithelial iris cyst


Fig. 11.17
Primary stromal iris cyst containing a fluid–debris level

particularly those arising from the pigment epithelium, are stationary and asymptomatic.

1. **Epithelial** cysts are unilateral, solitary, dark-brown, globular structures which transilluminate. They may be located at the pupillary border (Fig. 11.14), in the mid-zone (Fig. 11.15) or the iris root. Occasionally the cysts become dislodged and float freely in the anterior chamber or vitreous. The vast majority are innocuous and rarely require treatment.
2. **Stromal** cysts present in the first year of life. They are solitary, unilateral, have a smooth, translucent anterior wall and contain fluid (Fig. 11.16). The cyst may remain dormant for many years or suddenly enlarge and cause secondary glaucoma, corneal decompensation and show a fluid–debris level reminiscent of a pseudo-hypopyon (Fig. 11.17). Occasionally the cyst may break free from the iris and float in the anterior chamber or migrate to another location. Although spontaneous regression can occur, most require treatment by needle aspiration or surgical excision.


Fig. 11.18
Secondary iris cyst containing a worm

Secondary cysts

Secondary iris cysts develop as a result of implantation, parasites (Fig. 11.18), tumours or the prolonged use of

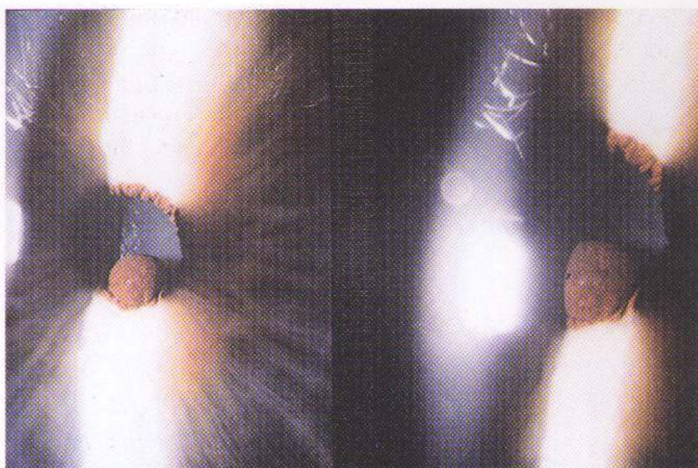


Fig. 11.19
Secondary iris cysts due to topical administration of long-acting miotics

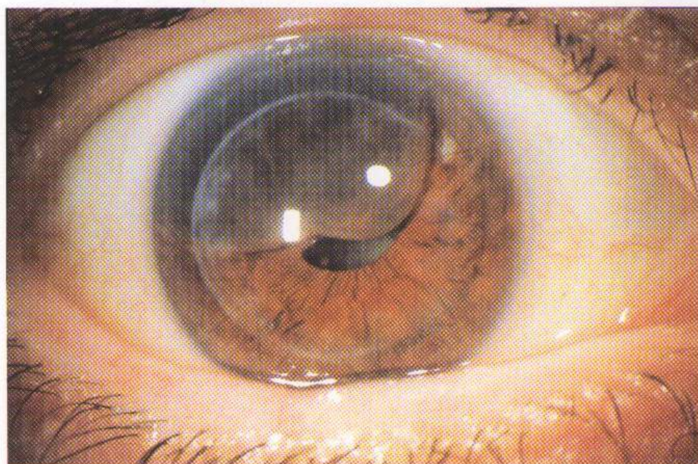


Fig. 11.20
Secondary iris cyst following corneal grafting

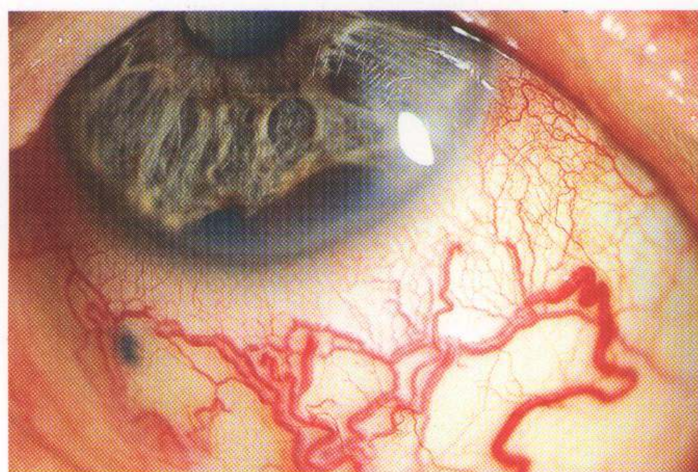


Fig. 11.21
Sentinel vessels associated with ciliary body melanoma
(Courtesy of B. Damato)

long-acting miotics. The latter are usually bilateral, small, multiple and located along the pupillary border (Fig. 11.19). Their development can be prevented by the use of topical 2.5% phenylephrine. Implantation cysts originate by deposition of surface epithelial cells from the conjunctiva or cornea on the iris after penetrating or surgical trauma and may lead to the following:

1. **Pearl** cysts are small, white, solid lesions with opaque walls located in the stroma and are not connected to the wound.
2. **Serous** cysts are translucent, filled with fluid and may be connected to the wound (Fig. 11.20). They frequently enlarge, leading to corneal oedema, anterior uveitis and glaucoma. Ultrasound biomicroscopy may show the location and extent of the lesions when surgical excision is contemplated.

Ciliary body tumours

Ciliary body melanoma

About 10% of uveal melanomas arise from the ciliary body.

Clinical features

1. **Presentation** is in the sixth decade with visual symptoms but occasionally the tumour may be discovered incidentally.
2. **Signs** depend on the size and location of the tumour. Small localized tumours cannot usually be visualized without pupillary dilatation and gonioscopy.
   - Dilated episcleral blood vessels in the same quadrant as the tumour (sentinel vessels) (Fig. 11.21).
   - Extraocular extension through the scleral emissary vessels may produce a dark epibulbar mass (Fig. 11.22) which may be mistaken for a conjunctival melanoma.
   - Pressure on the lens may give rise to astigmatism, subluxation or cataract (Fig. 11.23).
   - Erosion through the iris root may mimic iris melanoma (Fig. 11.24).
   - Retinal detachment may be caused by posterior extension.



Fig. 11.22
Extraocular extension of ciliary body melanoma

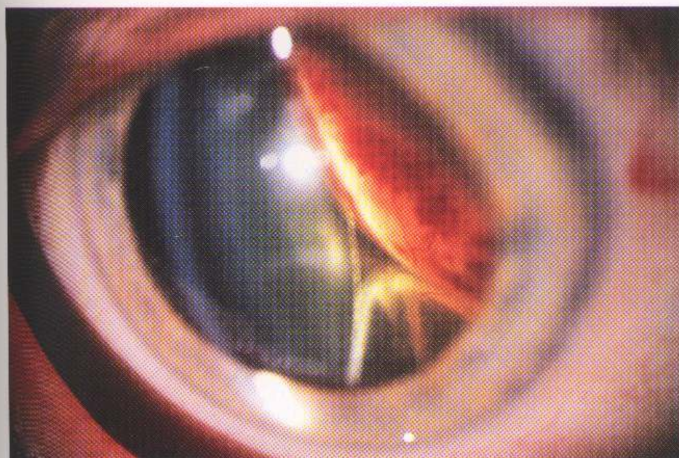


Fig. 11.23
Ciliary body melanoma displacing the lens (Courtesy of C. Barry)

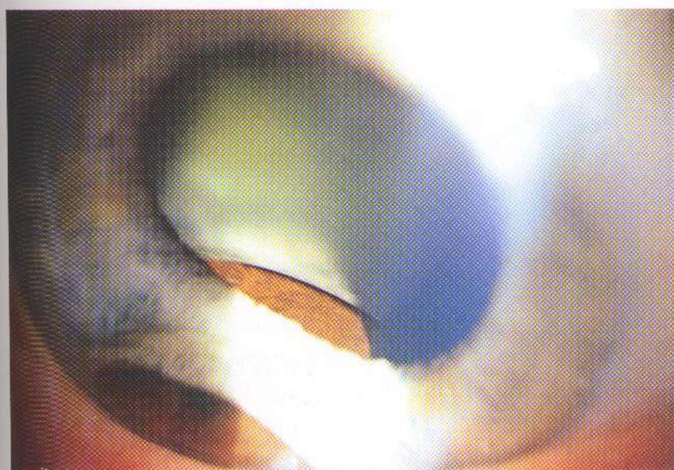


Fig. 11.24
Ciliary body melanoma eroding the iris root

- Anterior uveitis, caused by tumour necrosis, is uncommon.
- Circumferential (annular) growth for 360° carries the worst prognosis because early diagnosis is difficult.

Investigations

1. **Triple-mirror contact lens** examination through a well-dilated pupil is essential and is particularly useful in detecting forward erosion through the iris root into the angle.
2. **Transillumination** may give an approximate indication of tumour extent but is of little diagnostic value because an amelanotic melanoma may transilluminate.
3. **Ultrasonic biomicroscopy** is useful in eyes with opaque media and also shows dimensions and extent.
4. **Biopsy** may be helpful in selected cases.

Treatment

1. **General considerations** (*see* choroidal melanoma below).
2. **Enucleation** for large tumours and those affecting the anterior choroid. Secondary glaucoma, resulting from extensive invasion of Schlemm canal, is also an indication for enucleation.
3. **Iridocyclectomy** for small or medium-sized tumours involving no more than one-third of the angle. Complications are vitreous haemorrhage, retinal detachment and incomplete resection.
4. **Radiotherapy** by brachytherapy or external beam irradiation in selected cases.

Differential diagnosis

1. **Uveal effusion syndrome** (*see* Fig. 12.66) may resemble circumferential ciliary body melanoma. However, the effusion is lobulated, transilluminates brightly and appears cystic on ultrasonography.
2. **Congenital epithelial iridociliary cysts** may displace the lens but can be readily differentiated from melanomas by ultrasonography.
3. **Other ciliary body tumours**, which are extremely rare, include medulloepithelioma, metastases, adenocarcinoma, cystic adenoma and leiomyoma. In most of these the correct diagnosis can be made only histologically.

Choroidal tumours

Choroidal melanoma

Melanoma of the choroid is the most common primary intraocular tumour in adults and accounts for 85% of uveal melanomas.

Clinical features

1. **Presentation** is usually during the sixth decade (range fifth to eighth) in one of the following ways:

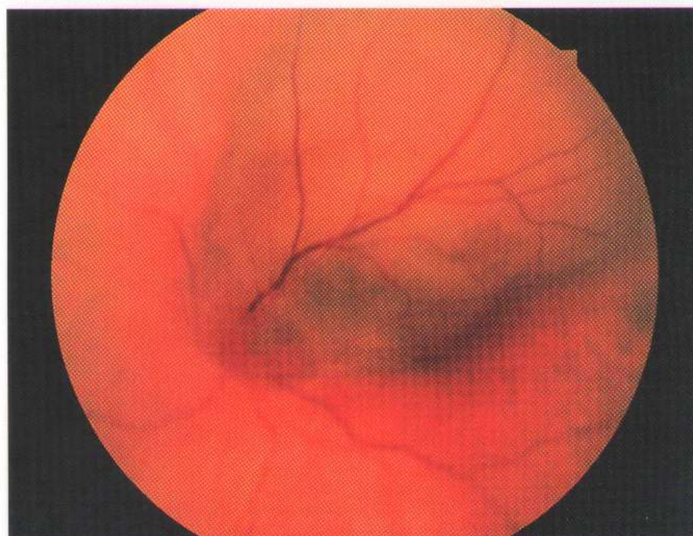


Fig. 11.25
Choroidal melanoma

- An asymptomatic tumour detected by chance.
- A symptomatic tumour causing decreased visual acuity or a visual field defect.
- About a third of patients complain of very brief 'balls of light' travelling across the visual field two to three times a day, most apparent in subdued lighting.

2. Signs

- An elevated, subretinal, dome-shaped, brown or grey mass (Fig. 11.25). Occasionally the tumour may be mottled with dark-brown or black pigment, or be virtually amelanotic (Fig. 11.26). Surface orange pigment (lipofuscin) is common but not diagnostic.
- If the tumour breaks through Bruch membrane it acquires a mushroom-shaped appearance (Fig. 11.27).

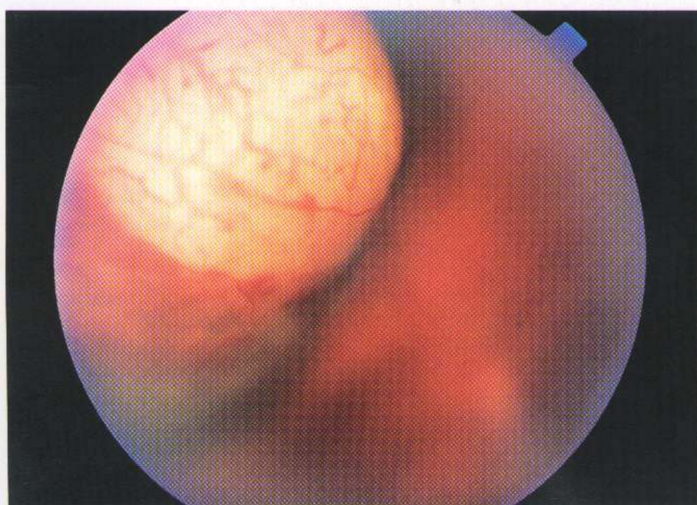


Fig. 11.26
Amelanotic choroidal melanoma

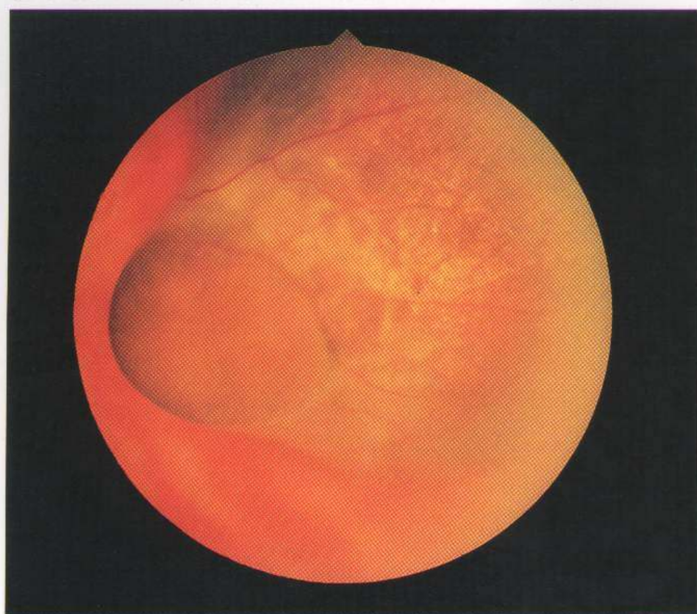


Fig. 11.27
Mushroom-shaped choroidal melanoma with surface lipofuscin (Courtesy of D. Lehman)

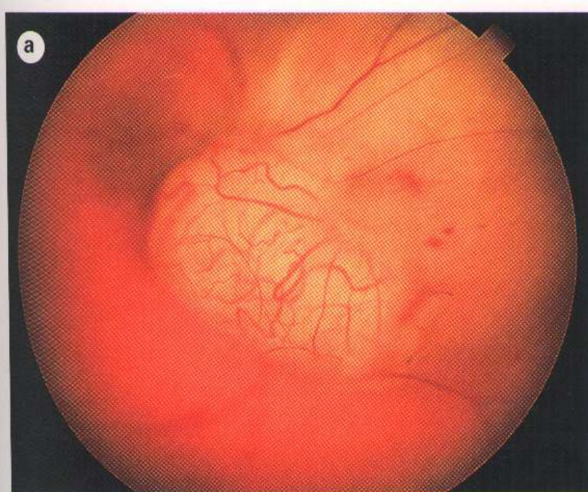


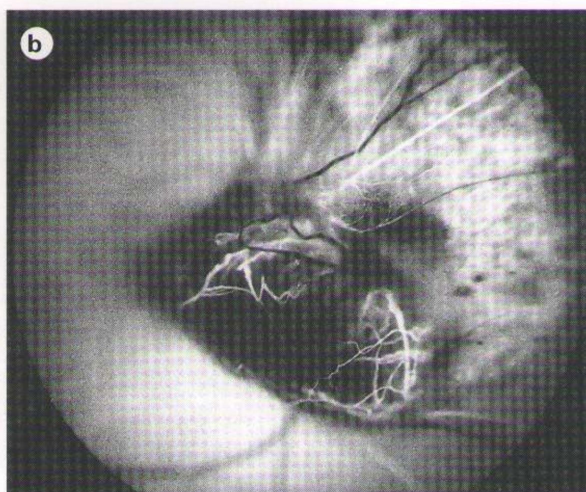
Fig. 11.28
B-scan ultrasonogram showing a choroidal melanoma and secondary retinal detachment

- A secondary exudative detachment is common and must not be mistaken for a rhegmatogenous detachment.
- Occasional features include choroidal folds, haemorrhage, secondary glaucoma, cataract and uveitis.

Investigations

1. **Binocular indirect ophthalmoscopy** is sufficient for accurate diagnosis in the vast majority of cases.
2. **Indirect slit-lamp biomicroscopy** detects subtle features associated with relatively small tumours, such as the presence of lipofuscin pigment, subretinal fluid, cystoid changes in the overlying sensory retina and dilated vessels within the tumour.
3. **Ultrasonography** is used to determine tumour size and detect extraocular extension. B-scan ultrasonography shows the anterior border of the tumour as well as acoustic hollowness, choroidal excavation and orbital shadowing (Fig. 11.28).
4. **FA** is of limited diagnostic value because there is no pathognomonic pattern. Most melanomas show a 'dual circulation' (Fig. 11.29b and c), mottled fluorescence during the arteriovenous phase and progressive leakage and staining (Fig. 11.29d).
5. **ICG** is superior to FA because there is less interference caused by RPE changes, better visualization of the tumour and choroidal vessels, and superior definition of tumour margins.
6. **MRI**, particularly when combined with surface coils and fat suppression sequences, shows that choroidal melanomas are hyperintense in T1-weighted (Fig. 11.30) and hypointense in T2-weighted images, but these features are not pathognomonic.
7. **Colour-coded Doppler** imaging may differentiate pigmented tumours from intraocular haemorrhage, particularly in eyes with opaque media.
8. **Fine-needle aspiration biopsy** is used occasionally to obtain cellular aspirates for analysis where the diagnosis cannot be established by less invasive methods.





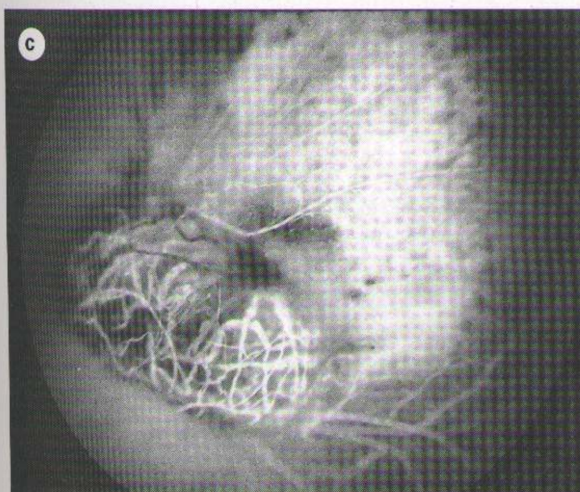




Fig. 11.29
(**a**) Choroidal melanoma; (**b** and **c**) FA early phases showing a 'dual circulation'; (**d**) late phase showing mild hyperfluorescence due to leakage (Courtesy of S. Milewski)

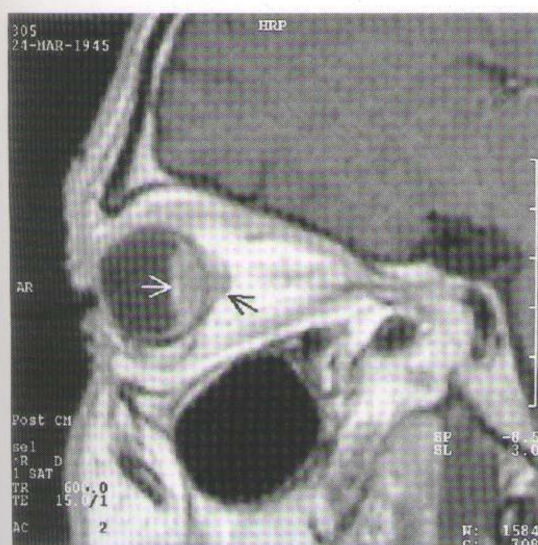


Fig. 11.30
T1-weighted MRI scan showing a choroidal melanoma (white arrow) and extraocular extension (black arrow) (Courtesy of M. Karolczak-Kulesza)

9. **General medical examination** is aimed at:
   - Excluding a metastasis *to* the choroid, which occurs most frequently from the bronchus in both sexes and the breast in women. Occasionally, the primary site is the kidney or gastrointestinal tract. Initial investigations should include chest radiography, rectal examination and mammography in females.
   - Detecting metastatic spread *from* the choroid as this would influence management. For example, a patient with overt metastatic disease would not be subjected to enucleation of a painless eye. The liver is by far the most frequent site for metastases. Hepatic involvement can be detected by ultrasonography and elevated gamma-glutamyl transpeptidase and alkaline phosphatase levels. Chest radiography should also be performed to detect possible lung secondaries but these are rare without liver metastases.

Treatment

This is complex and should be tailored to the individual patient. The following factors are considered: (a) size, location, extent and apparent activity of the tumour, (b) state of the fellow eye, (c) general health and age of the patient and (d) the patient's wishes and fears. For instance, treatment may be inappropriate for a slowly growing tumour in the only eye of a very elderly or chronically ill patient. There is an increasing trend towards the use of combined therapy with the following modalities:

1. **Brachytherapy** is frequently the treatment of first choice because it is relatively straightforward and effective.
   a. *Indications* are tumours less than 10 mm in elevation and less than 20 mm in basal diameter in which there is a reasonable chance of salvaging vision. Supplemental transpupillary thermotherapy may be required to enhance the results. Tumour regression starts about 1–2 months after treatment and continues for several years.
   b. *Complications* include retinopathy, papillopathy, vitreous haemorrhage, cataract and recurrence of tumour.
   c. *Cure rate* is similar to that following enucleation of comparable tumours.
2. **External radiotherapy** with protons or helium ions by means of a cyclotron unit. The advantages over brachytherapy are that the beam can be more highly focused, affording a more homogeneous dose of radiation. Treatment is fractionated over 4 days, with each dose delivered over a 30-second period.
   a. *Indications* are tumours unsuitable for brachytherapy either because of size or posterior location to within 4 mm of the disc or fovea.
   b. *Complications*, which are more common following treatment of large tumours, include loss of lashes, eyelid depigmentation, canaliculitis with epiphora, conjunctival keratinization, keratitis, exudative retinal detachment and neovascular glaucoma.
   c. *Cure rate* is similar to that following brachytherapy or enucleation.
3. **Transpupillary thermotherapy** (TTT) is performed with a diode laser to induce hyperthermia but not coagulation.
   a. *Indications* are selected small tumours, particularly if pigmented and located near the fovea or optic disc. TTT is also a complementary modality to brachytherapy.
   b. *Complications* include visual field defects and maculopathy.
4. **Trans-scleral local resection** is a difficult procedure which involves excision of the tumour with a rim of healthy choroid under a partial-thickness scleral flap. The procedure is performed under systemic arterial hypotension.
   a. *Indications* are carefully selected tumours that are too thick for radiotherapy and usually less than 16mm in diameter.
   b. *Complications* include haemorrhage, retinal detachment, cataract and tumour recurrence.
5. **Stereotactic radiosurgery** with the Gamma-knife is a new method involving single-session delivery of ionizing radiation to a stereotactically localized volume of tissue. This modality is an alternative to charged particle irradiation or enucleation in treating large tumours with preservation of visual function in selected cases.
6. **Enucleation**
   a. *Indications* for excision of the globe are very large tumours, particularly if all useful vision has been irreversibly lost. In these cases, enucleation is preferred to brachytherapy because the dose of radiation required to reach the apex of the tumour would be too great to salvage the rest of the globe.
   b. *Technique* should be meticulous to avoid dissemination of malignant cells. The procedure should be performed with gentle isolation and section of the extraocular muscles, and minimal traction on the optic nerve when cutting. About 4 weeks after enucleation, the patient can be fitted with an artificial eye.
7. **Exenteration** of the orbit is indicated for patients with extensive extraocular extension at the time of diagnosis or for orbital recurrences following enucleation.
8. **Palliation** with chemotherapy and/or immunotherapy may prolong life in patients with metastatic disease. In patients with lung metastases, life expectancy is generally <1 year and, when the liver is involved, <6 months.

Modified Callender classification of uveal melanomas

1. **Spindle cell melanomas**, which make up 45% of all tumours, are composed of spindle cells, with a small proportion described as fascicular because of the palisading or ribbon-like arrangement of cells in parallel rows (Fig. 11.31).
2. **Pure epithelioid cell melanomas** make up 5% (Fig. 11.32).
3. **Mixed cell melanomas**, containing spindle and epithelioid cells, account for 45% (Fig. 11.33).
4. **Necrotic melanomas**, in which the predominant cell type is unrecognizable, make up the remaining 5% (Fig. 11.34).

Prognostic factors

1. **Histological features** implying an adverse prognosis include large numbers of epithelioid cells per high-power field, closed vascular loops within the tumour and lymphocytic infiltration.

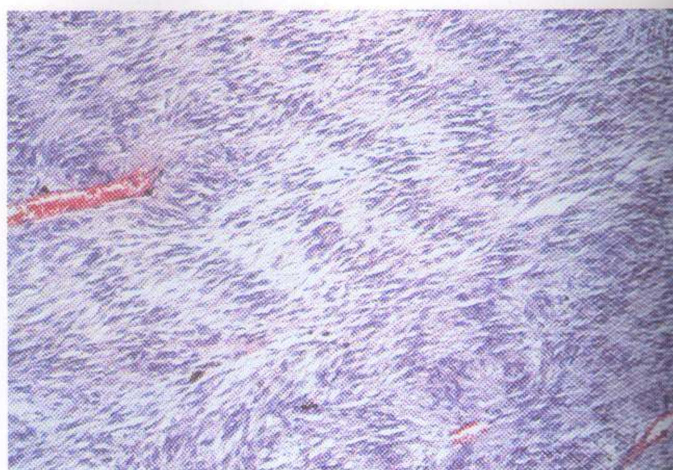
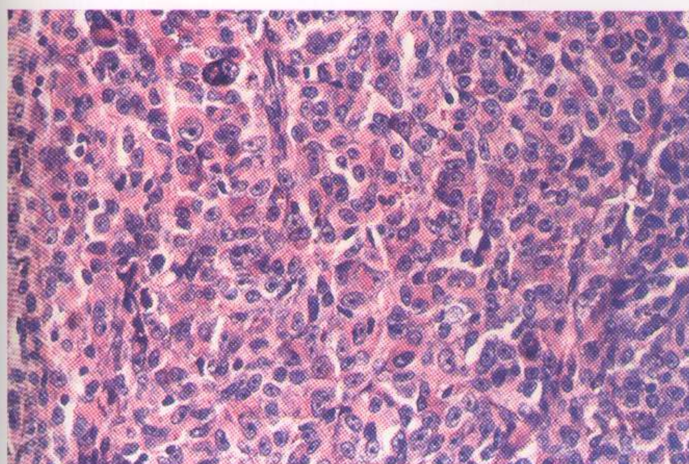
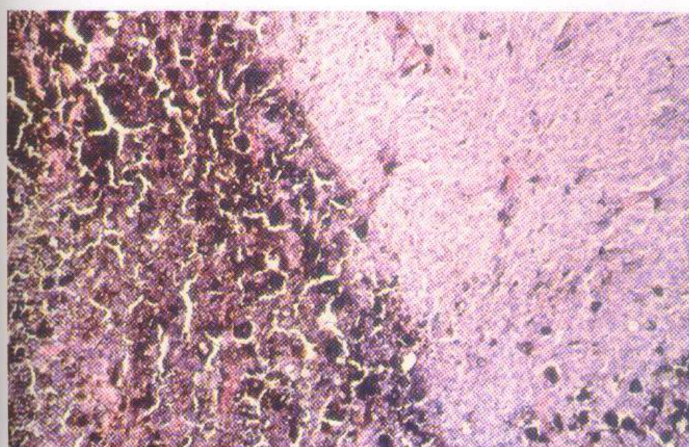
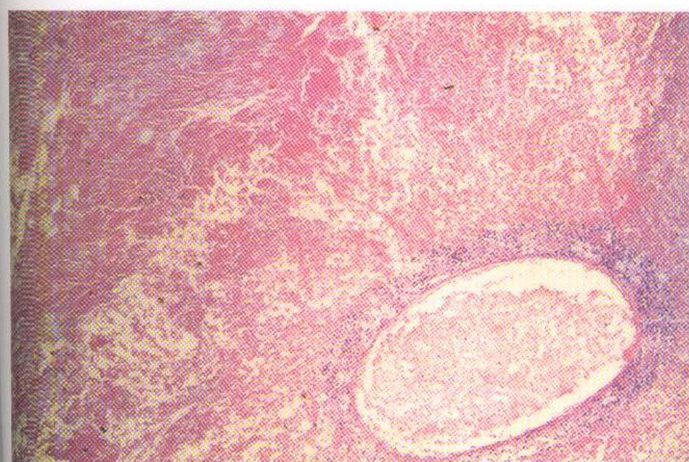


Fig. 11.31
Fascicular spindle cell melanoma (Courtesy of A. Garner)


Fig. 11.32
Epithelioid cell melanoma (Courtesy of A. Garner)


Fig. 11.33
Mixed cell melanoma (Courtesy of A. Garner)


Fig. 11.34
Necrotic melanoma (Courtesy of A. Garner)

2. **Chromosomal abnormalities** within the melanoma cells are associated with a very poor prognosis, with a 50% death rate at 5 years.
3. **Large tumours** have a worse prognosis than small tumours. Following enucleation, 5-year mortality is as follows:
   - Small tumours (<10 mm in diameter): 16%.
   - Medium tumours (10–15 mm in diameter): 32%.
   - Large tumours: 53%.
4. **Extrascleral extension** carries a very poor prognosis.
5. **Location**: anterior tumours have a worse prognosis because they are usually diagnosed later than those near the posterior pole.
6. **Patients over the age of 65 years** have a worse prognosis than younger patients.

Differential diagnosis

Although in the vast majority of cases the diagnosis is straightforward, the following conditions should be considered in the differential diagnosis of atypical cases, particularly amelanotic melanomas:

1. **Other choroidal tumours**, most notably large naevi, circumscribed haemangiomas and solitary metastases.
2. **Solitary choroidal granulomas** associated with sarcoidosis or tuberculosis.
3. **Posterior scleritis** (*see* Chapter 7).

Choroidal naevus

Choroidal naevi are present in about 5% of the general population but are less common in fair-skinned individuals. Although they are probably present at birth, growth occurs mainly during the pre-pubertal years and is extremely rare thereafter. For this reason the rare event of clinically detectable growth should arouse suspicion of malignant transformation.

Signs

1. **Typical naevus**
   - An asymptomatic, oval or circular, slate-blue or green-grey lesion with detectable but not sharp borders which may be associated with surface drusen (Fig. 11.35 and *see* Fig. 11.38a).
   - Dimensions are <5 mm in diameter and 1 mm or less in thickness.
2. **A suspicious naevus** has one or more of the following characteristics:
   - Presence of symptoms, such as metamorphopsia or photopsia.
   - Dimensions >5 mm in diameter and >1 mm in thickness (Fig. 11.36).
   - Clumps of surface orange (lipofuscin) pigment (Fig. 11.37).
   - Absence of surface drusen on a thick lesion.
   - Location of the posterior margin of the lesion within 3 mm of the optic disc.
   - Serous retinal detachment either over the surface of the lesion or inferiorly.

NB: The greater the number of these features, the higher the chance that the lesion is a melanoma.

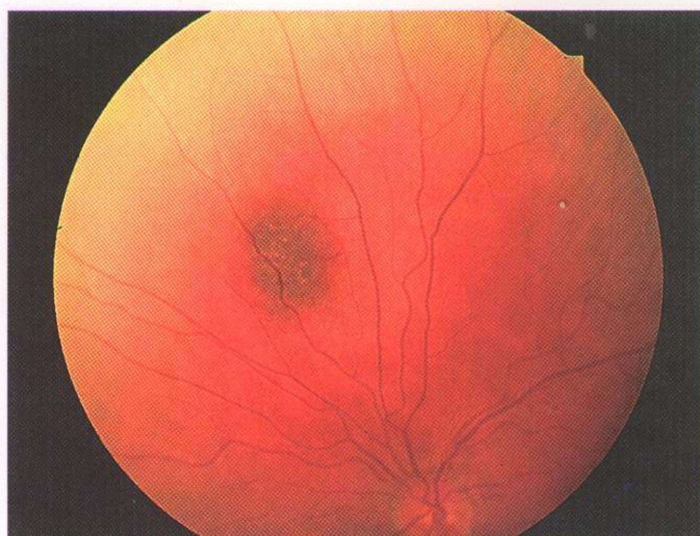


Fig. 11.35
Typical choroidal naevus

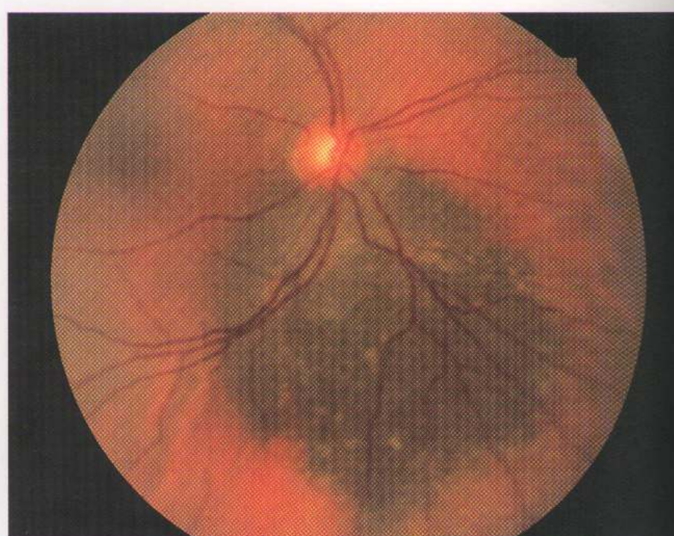


Fig. 11.36
Suspicious choroidal naevus

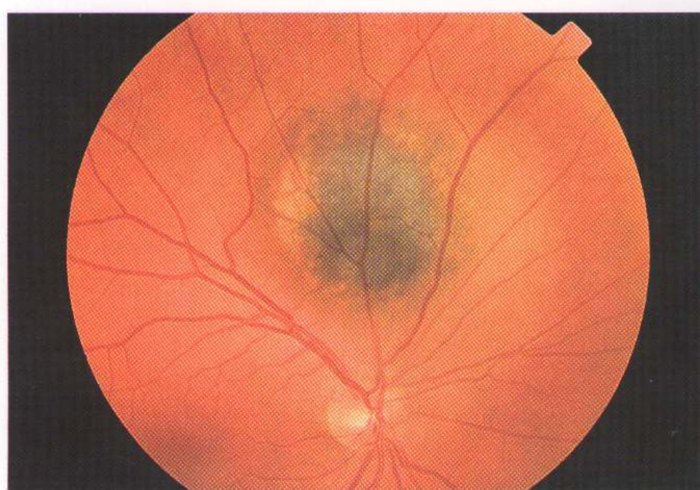
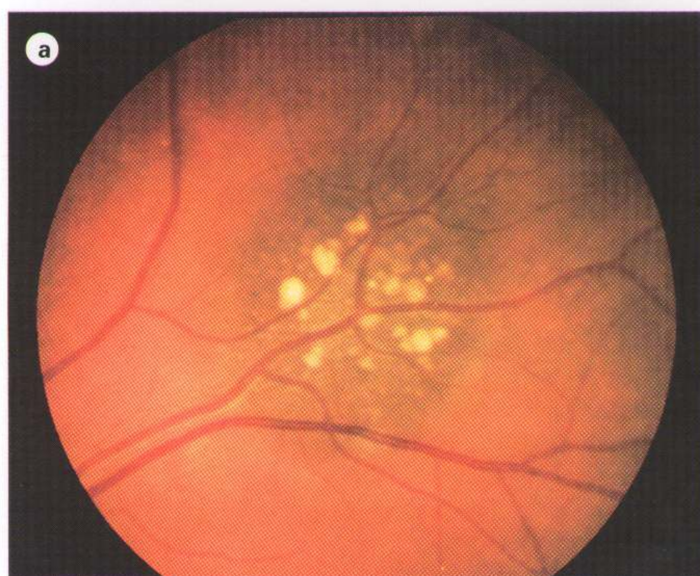


Fig. 11.37
Suspicious choroidal naevus with surface lipofuscin

Investigations

1. **FA** findings depend on the amount of pigmentation within the naevus and associated changes in the overlying RPE. Most choroidal naevi are avascular and pigmented, giving rise to hypofluorescence caused by blockage of background choroidal fluorescence. If the naevus is associated with surface drusen, this will result in areas of hyperfluorescence (Fig. 11.38b). FA is, however, not helpful in distinguishing a small melanoma from a naevus.
2. **Ultrasonography** shows a localized flat or slightly elevated lesion with high internal reflectivity (Fig. 11.39).



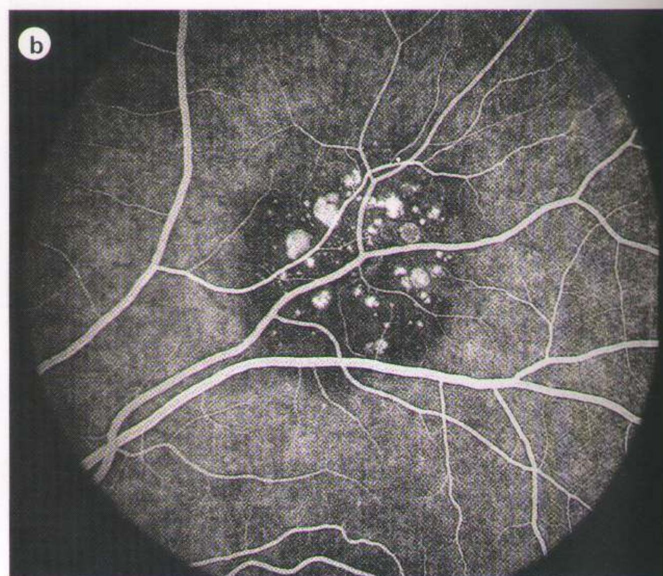


Fig. 11.38
(**a**) Typical choroidal naevus with surface drusen; (**b**) FA showing blockage of background fluorescence and hyperfluorescence of surface drusen



Fig. 11.39
B-scan ultrasonogram of choroidal naevus (*see text*) (Courtesy of M. Karolczak-Kulesza)

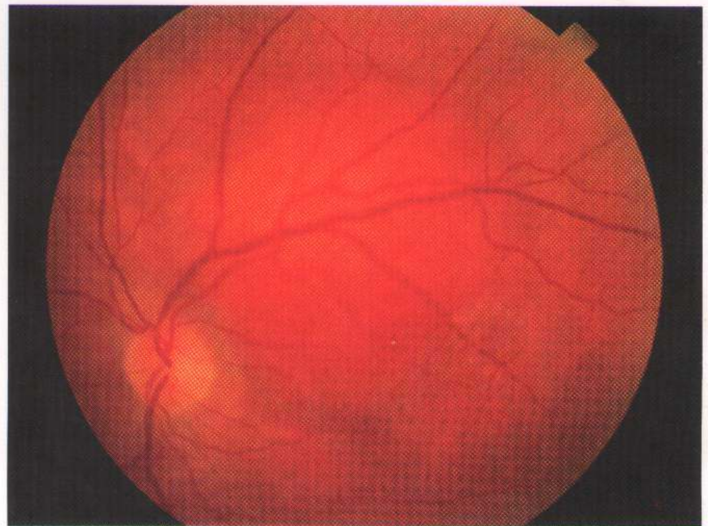


Fig. 11.40
Circumscribed choroidal haemangioma (Courtesy of J.A. Shields and A. Singh)

Management

1. **Typical naevi** do not require follow-up because the risk of malignant transformation is extremely low.
2. **Suspicious naevi** should be evaluated initially every 3–6 months and then 9–12 months with fundus photography and ultrasonography to detect the possibility of growth. Although it is difficult to detect small changes in thickness by ultrasonography, careful comparison of fundus photographs, with special attention to the location of landmarks such as blood vessels, is usually a reliable method of documenting growth. Once growth has been documented, the lesion should be reclassified as a choroidal melanoma and managed accordingly.

Circumscribed choroidal haemangioma

Circumscribed choroidal haemangioma is a rare, benign, vascular hamartoma which is probably present at birth but does not become symptomatic until years later. The tumour is almost always solitary and is not associated with systemic disease. Most are stationary and a few are slow-growing.

Clinical features

1. **Presentation** is in the fourth to fifth decades with unilateral central visual impairment.
2. **Signs**
   - A dome-shaped or placoid, orange-red, mass which blends with the surrounding choroid (Fig. 11.40). Subtle white foci may be present on the surface of the tumour and probably represent fibrous metaplasia of the overlying RPE.
   - The lesion usually measures 3–9 mm in diameter and is most commonly located in the juxtapapillary or macular area.
3. **Complications** include secondary cystoid retinal degeneration overlying the tumour, exudative retinal detachment, macular invasion, RPE degeneration and subretinal fibrosis.

Special investigations

1. **Ultrasonography** shows an oval or placoid lesion with a sharp anterior border and high internal reflectivity, but no choroidal excavation or orbital shadowing (Fig. 11.41).
2. **FA** reveals filling during the choroidal phase (Fig. 11.42a), progressive hyperfluorescence during the venous phase and late leakage (Fig. 11.42b).

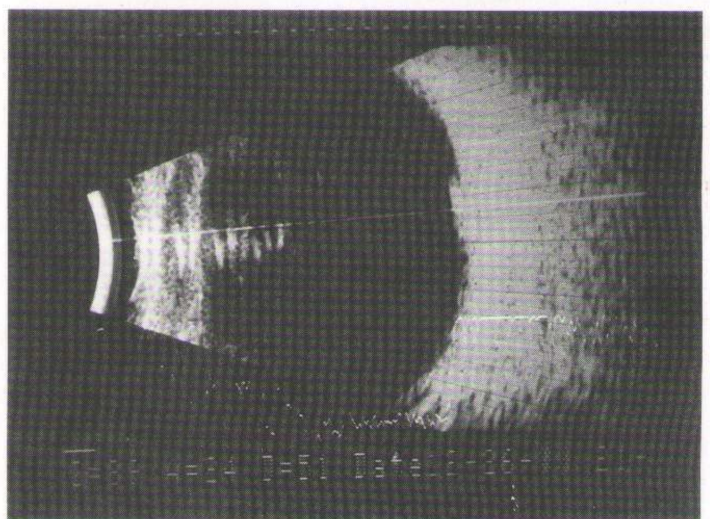
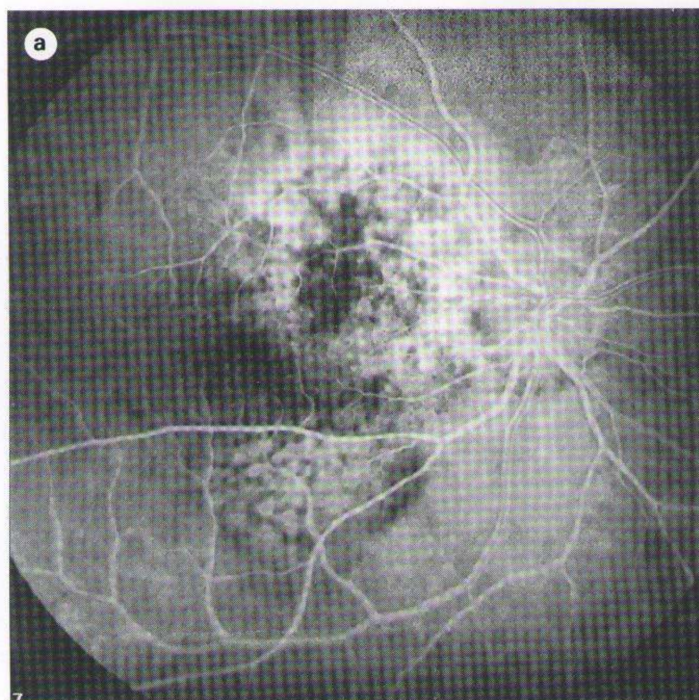


Fig. 11.41
B-scan ultrasonogram of circumscribed choroidal haemangioma (*see text*) (Courtesy of S. Milewski)



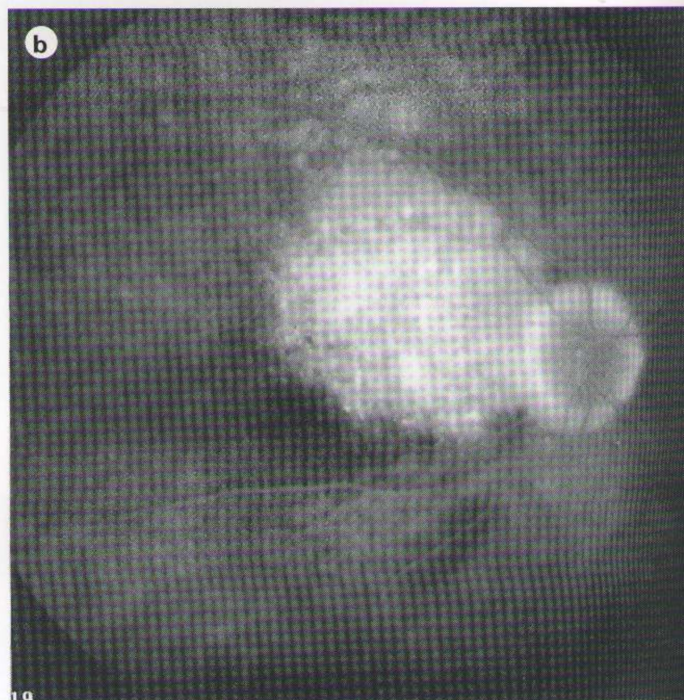


Fig. 11.42
FA of circumscribed choroidal haemangioma. (**a**) Early phase showing hyperfluorescence due to filling; (**b**) late phase showing leakage (Courtesy of S. Milewski)

Treatment

Vision-threatening lesions may be treated as follows:

1. **Transpupillary thermotherapy** for lesions not involving the macula. Indocyanine green can be administered as an adjunct to enhance uptake of diode laser energy.
2. **Radiotherapy** either by low-dose lens-sparing external beam irradiation or brachytherapy.

Diffuse choroidal haemangioma

The diffuse choroidal haemangioma usually affects over half of the choroid and enlarges very slowly. It occurs in patients with the Sturge–Weber syndrome ipsilateral to the naevus flammeus (*see* Chapter 20).

1. **Presentation** is in the third decade with visual impairment.
2. **Signs**. Thickening of the choroid and a deep-red 'tomato ketchup' colour which is most marked at the posterior pole (Fig. 11.43).
3. **Complications** include secondary retinal cystoid degeneration and exudative retinal detachment.
4. **Treatment** is by external beam radiotherapy.

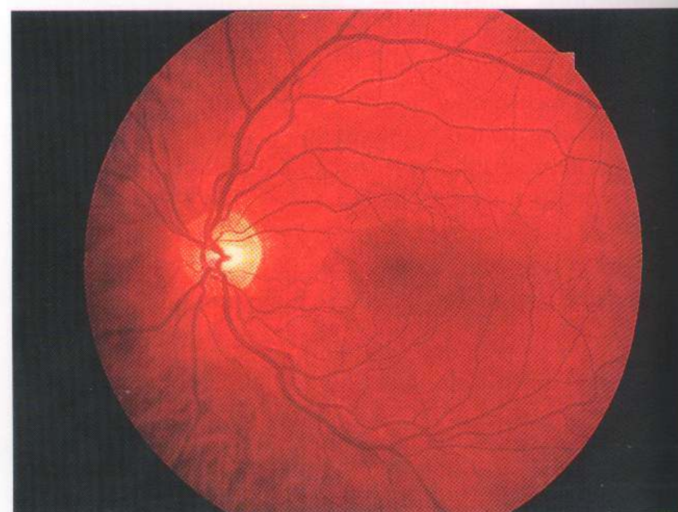


Fig. 11.43
Diffuse choroidal haemangioma

Metastatic tumours

The choroid is by far the most common site for uveal metastases, accounting for about 90%, followed by the iris and ciliary body. Metastatic tumours to the choroid are more common than primary malignancies but their presence is usually undetected or overshadowed by the patient's general illness. The most frequent primary site is the breast in women and the bronchus in both sexes. A choroidal secondary may be the initial presentation of a bronchial carcinoma, whereas a past history of breast cancer is the rule in patients with breast secondaries. Other less common primary sites include the gastrointestinal tract, kidney and skin melanoma. The prostate is, however, an extremely rare primary site. Patient survival is generally poor, with a median of 8–12 months for all patients and 15–17 months for those with breast carcinoma. In patients with breast carcinoma risk factors for choroidal metastases include dissemination of disease in more than one organ and the presence of lung and brain metastases.

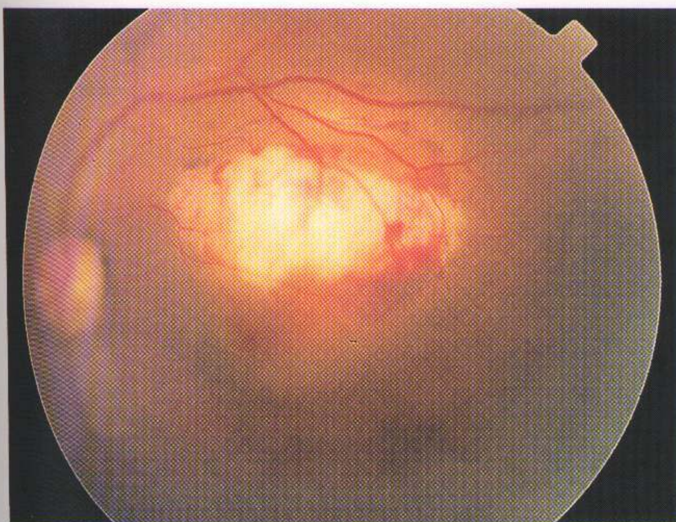


Fig. 11.44
Choroidal metastasis from colon carcinoma (Courtesy of S. Milewski)

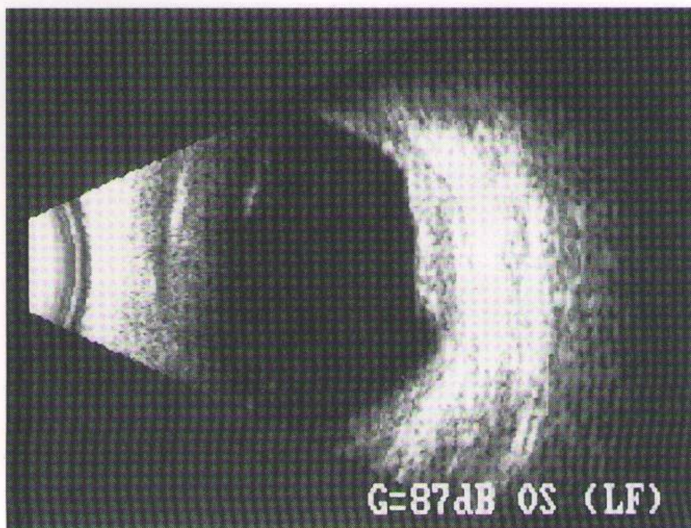


Fig. 11.46
Ultrasonogram of choroidal metastasis (*see text*) (Courtesy of M. Karolczak-Kulesza)

Clinical features

1. **Presentation** is usually with visual impairment although metastases may be asymptomatic if located away from the macula.
2. **Signs**
   - A fast-growing, creamy-white, placoid or oval lesion most frequently located at the posterior pole (Fig. 11.44).
   - The tumour seldom becomes significantly elevated because it infiltrates laterally (Fig. 11.45).
   - Occasionally the deposits assume a globular shape and may mimic an amelanotic melanoma.
   - The deposits may be multiple and both eyes are involved in 10–30% of cases.
   - Secondary exudative retinal detachment is frequent and may occur in eyes with relatively small deposits.

Special investigations

1. **Ultrasonography** shows diffuse choroidal thickening and moderate internal acoustic reflectivity (Fig. 11.46).
2. **FA** shows early hyperfluorescence (Fig. 11.47b) with diffuse late staining (Fig. 11.47c and d) but in contrast with choroidal melanomas a 'dual circulation' does not occur.

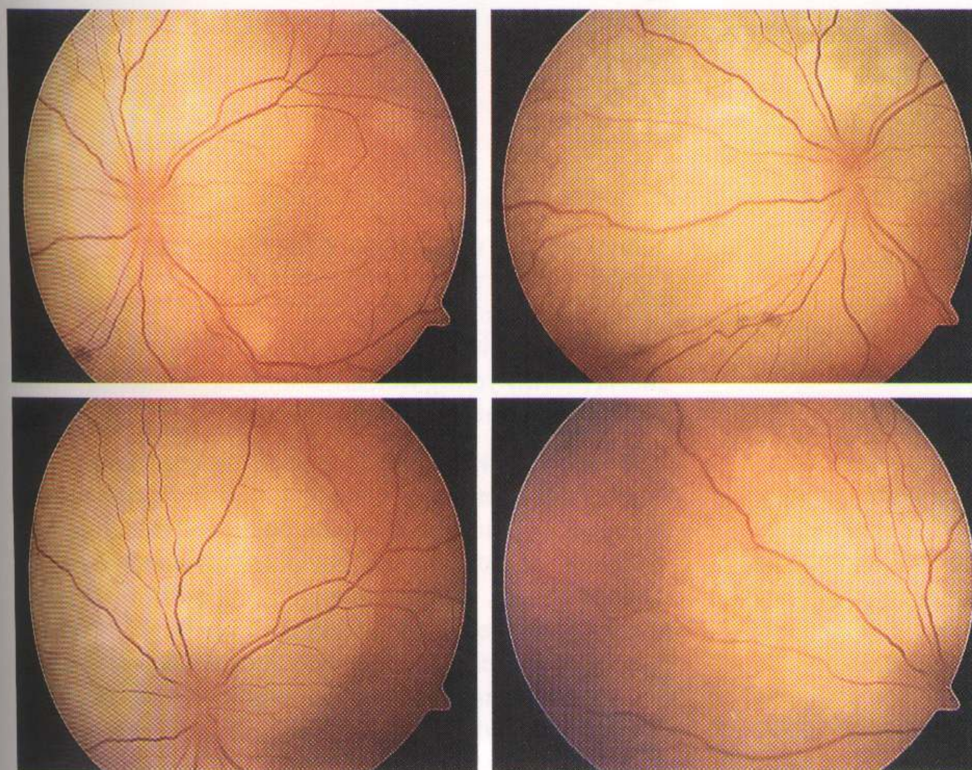


Fig. 11.45
Choroidal metastasis from breast carcinoma (Courtesy of M. Karolczak-Kulesza

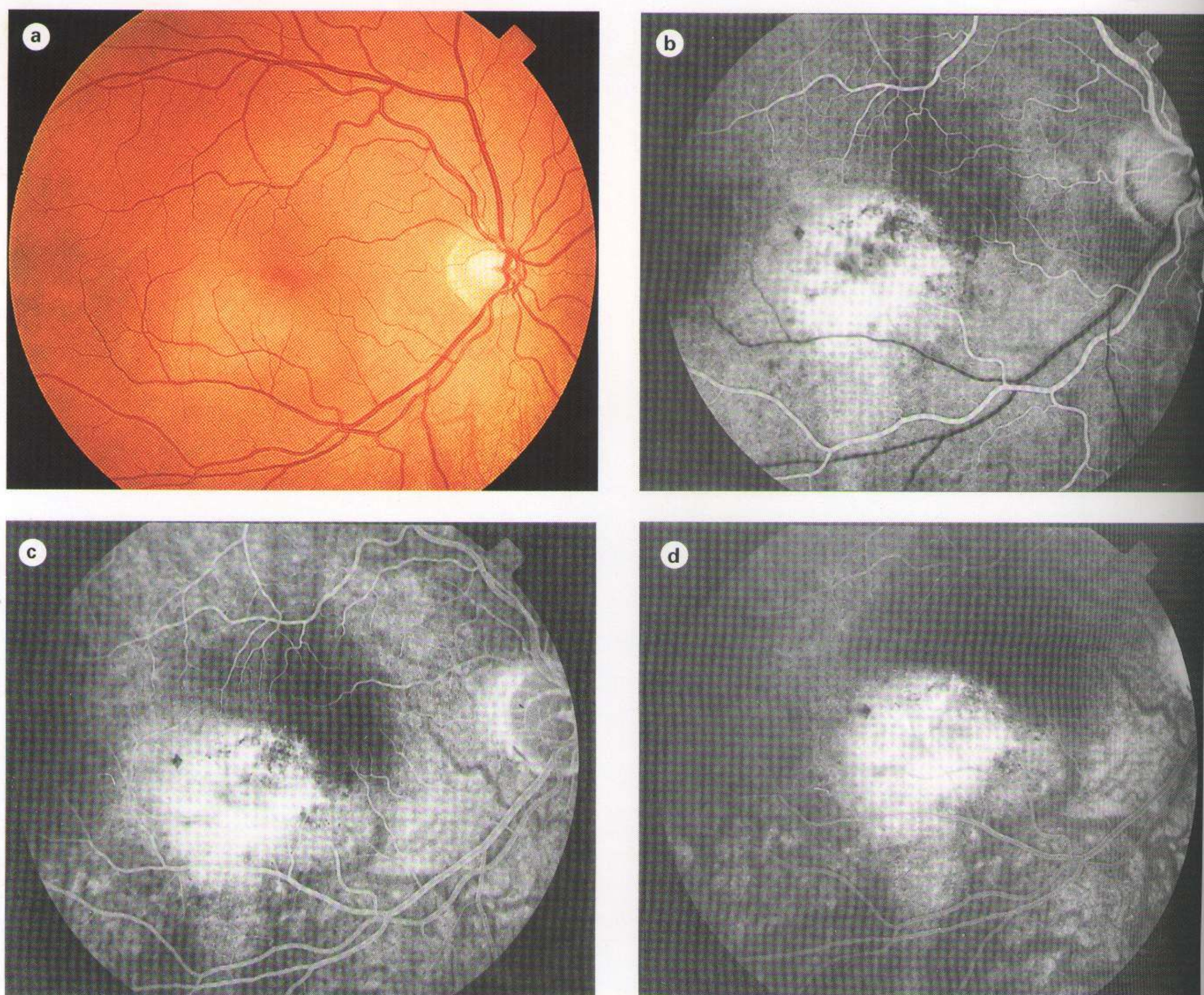


Fig. 11.47
(a) Choroidal metastasis; **(b)** FA arterial phase showing hyperfluorescence but absence of a 'dual circulation'; **(c, d)** late phases showing staining (Courtesy of S. Milewski)

3. **Biopsy**, performed either trans-sclerally or by fine-needle aspiration, may be useful.
4. **Systemic** investigations to detect the primary tumour, if unknown, or other metastatic sites.

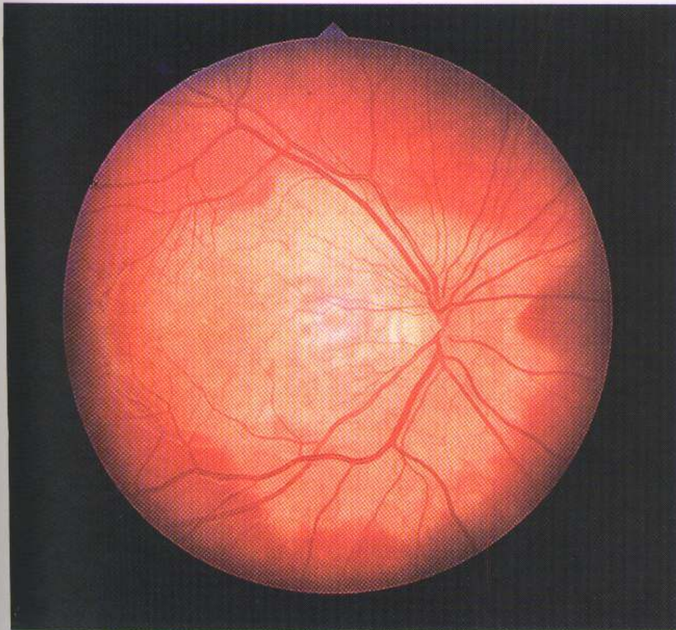
Treatment

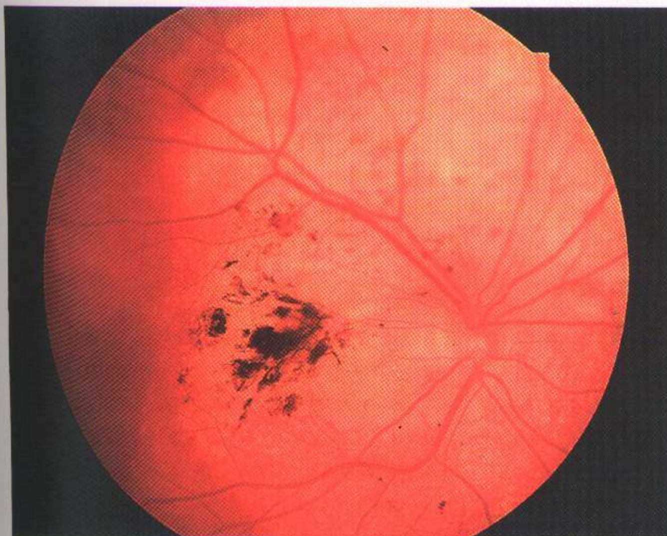
1. **Observation**, if the patient is asymptomatic or receiving systemic chemotherapy.
2. **Radiotherapy**, either external beam or brachytherapy, for small tumours.
3. **Transpupillary thermotherapy** is useful for deposits of moderate thickness and minimal amount of subretinal fluid.
4. **Systemic therapy** for the primary tumour may be beneficial for choroidal metastases.
5. **Enucleation** may be required for a painful blind eye.

Choroidal osseous choristoma

Choroidal osseous choristoma (osteoma) is a very rare, benign, very slow-growing, ossifying tumour which typically affects healthy young women. Both eyes are affected in about 25% of cases but not usually simultaneously.

1. **Presentation** is in the second to third decades with gradual visual impairment if the macular is involved.
2. **Signs**. An orange-yellow lesion with well-defined scalloped borders, most commonly situated at the posterior pole (Fig. 11.48). The tumour grows very slowly with the development of overlying RPE changes (Fig. 11.49).
3. **Complications**. Secondary choroidal neovascularization is common and responds poorly to laser photocoagulation.
4. **FA** shows a diffuse mottled pattern of hyperfluorescence during the early and late phases.

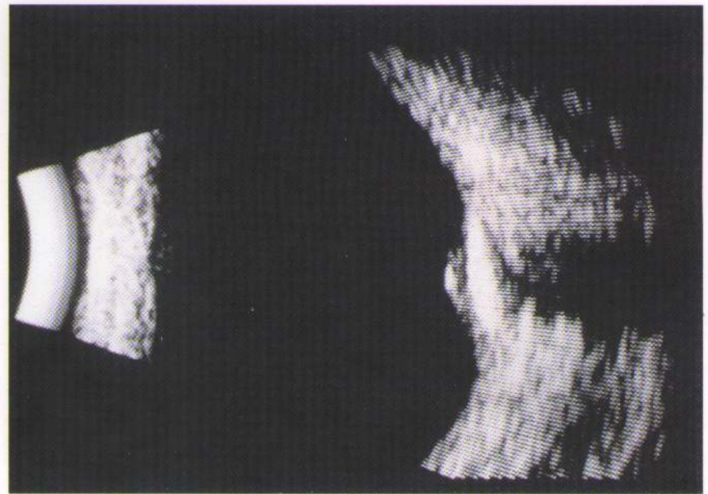

Fig. 11.48
Choroidal osseous choristoma

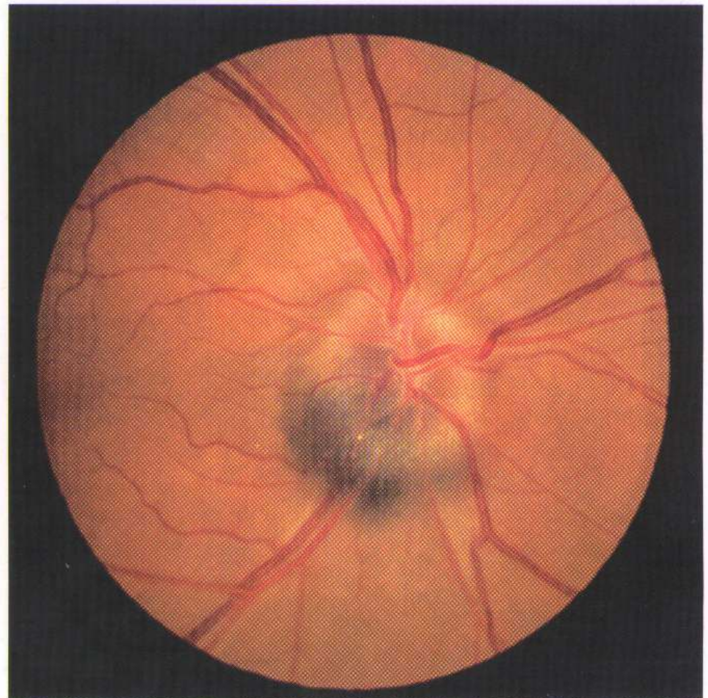

Fig. 11.49
Same eye several years later showing overlying RPE changes

5. **Ultrasonography** shows a very dense highly reflective lesion (bone) which renders silent the orbital tissue behind it (Fig. 11.50).

Melanocytoma

Melanocytoma is a benign, heavily pigmented tumour that may occur anywhere in the uveal tract, but arises most frequently from dendritic uveal melanocytes in the lamina cribrosa of the optic nerve head. Anterior uveal melanocytomas may undergo acute necrosis with resultant uveitis, pigment dispersion and secondary glaucoma. In contrast with choroidal melanoma, melanocytoma typically affects dark-skinned individuals, although it may also occur in white people.


Fig. 11.50
Ultrasonogram of choroidal osseous choristoma (*see text*)


Fig. 11.51
Melanocytoma of the optic nerve head

1. **Presentation** of optic nerve head melanocytoma is usually by chance, although a deep-seated tumour may cause optic nerve dysfunction.
2. **Signs**. A black lesion with feathery edges frequently occupying the inferior part of the disc (Fig. 11.51). Occasionally, the tumour is elevated and occupies the entire disc surface.
3. **Complications**, which are rare, include malignant transformation and central retinal vascular obstruction secondary to spontaneous tumour necrosis.
4. **Treatment** is not required except in the very rare event of malignant transformation.
5. **Differential diagnosis** includes choroidal melanoma invading the optic nerve head and reactive RPE hyperplasia.

Lymphoma

Primary intraocular–central nervous system lymphoma is an uncommon, diffuse, highly malignant, large B-cell (non-Hodgkin) lymphoma. It arises within the brain, spinal cord, leptomeninges and/or the eye and has a poor prognosis, with a 5-year survival rate of less than 33%.

CNS features

1. **At presentation** the following four profiles are seen:
   - Solitary or multiple intracranial nodules.
   - Diffuse meningeal or periventricular lesions.
   - Localized intradural spinal masses.
   - Intraocular involvement.
2. **The diagnosis** is usually made by identifying malignant lymphocytes in the brain, cerebrospinal fluid or vitreous.

Ocular features

Lymphoma tends to involve the vitreous and retina and frequently represents a diagnostic challenge, masquerading as uveitis. Ocular findings usually precede CNS involvement by months or a few years and only 20% of patients have ocular lesions at the time of diagnosis of CNS disease. Both eyes are eventually affected in 80% of cases, but the severity of involvement is often asymmetrical.

1. **Chronic anterior uveitis** unresponsive to steroids.
2. **Intermediate uveitis** in an elderly patient may initially be responsive to steroids but subsequently becomes unresponsive. The vitreous typically shows large clumps or sheets composed of malignant cells.
3. **Posterior segment**
   - Multifocal, large, yellowish, sub-RPE infiltrates are most commonly seen (Fig. 11.52). Coalescence of the lesions may form a ring which is pathognomonic for intraocular lymphoma (Fig. 11.53).
   - Less frequent manifestations include diffuse retinal infiltrates resembling viral retinitis, vascular sheathing and occlusion, and multifocal, tiny deep white lesions which can be misdiagnosed as inflammatory.
4. **Investigations** include neurological evaluation, MRI, lumbar puncture and vitreous biopsy.

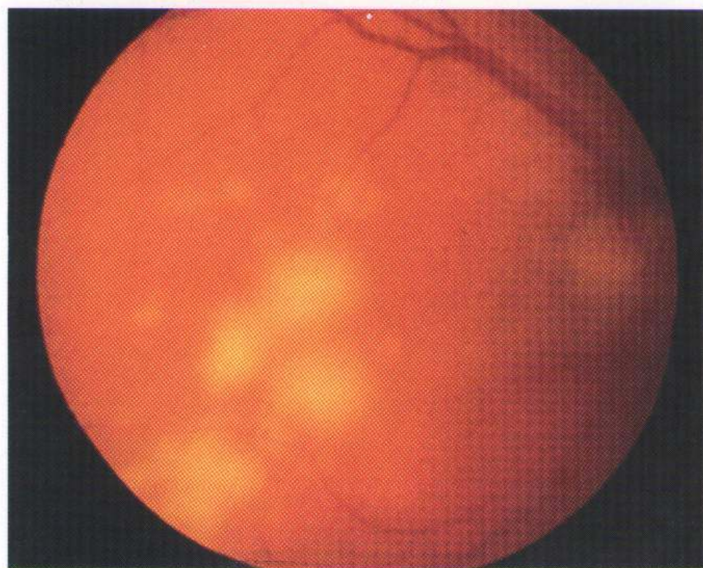


Fig. 11.52
Multifocal lymphomatous subretinal infiltrates (Courtesy of A. Cruess)

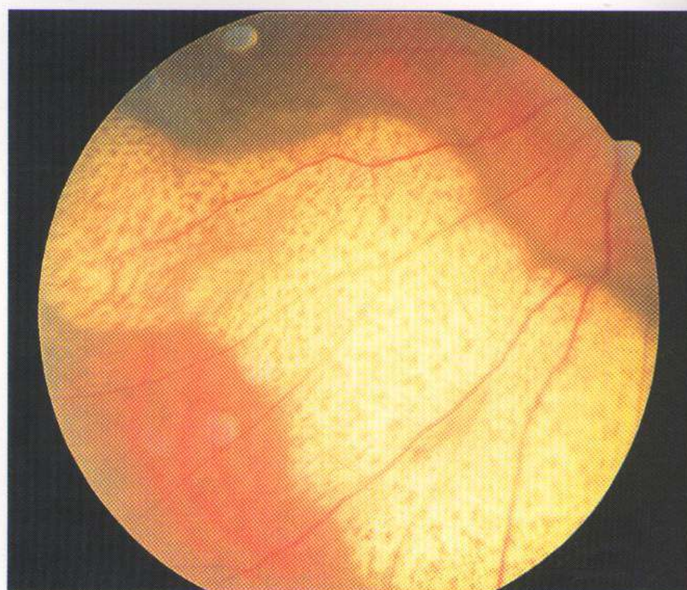


Fig. 11.53
Annular lymphomatous subretinal infiltration (Courtesy of B. Damato)

Treatment

1. **Systemic** treatment is with high-dose external beam radiotherapy to the eyes, sometimes in conjunction with whole brain radiotherapy, and/or systemic or intrathecal chemotherapy.
2. **Intravitreal** methotrexate may be used as primary treatment and for recurrences following systemic therapy.

Retinal and optic nerve head tumours

Retinoblastoma

Retinoblastoma is the most common primary intraocular malignancy of childhood. Even so, it is rare, occurring in about 1:20,000 live births and accounts for about 3% of all childhood cancers.

Genetics

Retinoblastoma results from malignant transformation of primitive retinal cells before final differentiation. Because these cells disappear within the first few years of life, the tumour is seldom seen after 3 years of age. Retinoblastoma

may be heritable or non-heritable. The predisposing gene (*RPE1*) is at 13q14.

1. **Heritable** (germline) retinoblastoma accounts for 40%. In these patients one allele of the *RPE1* (a tumour supressor gene) has mutated in all body cells. When a further mutogenic event ('second hit') affects the second allele, the cell undergoes malignant transformation. Because all the retinal precursor cells contain the initial mutation, these children develop bilateral and multifocal tumours. Familial cases also carry a predisposition to non-ocular cancers; most notably pinealoblastoma (trilateral retinoblastoma) and osteosarcoma. The risk of second malignancy increases greatly if external beam irradiation has been used to treat the original tumour, and the second tumour tends to arise within the irradiated field.
   - The risk of transmitting the gene mutation is 50%, and because of high penetrance 40% of offspring of a survivor of heritable retinoblastoma will develop the tumour.
   - Unaffected parents of a child with bilateral retinoblastoma with no family history have a 40% chance of producing another affected child.
   - Some familial cases present with initially unilateral disease and about 15% of patients with heritable retinoblastoma only express unilateral involvement.
2. **Non-heritable** (somatic) retinoblastoma accounts for 60% of cases. The tumour is unilateral, not transmissible and does not predispose the patient to an increased risk of second non-ocular cancers. Eighty-five per cent of patients with unilateral retinoblastoma fall into this category.

Presentation

The vast majority present within the first 2 years of life. Children with bilateral tumours tend to present earlier (average 12 months) than those with unilateral involvement.

1. **Leukocoria** (white pupillary reflex) is the commonest (60%) (Fig. 11.54).
2. **Strabismus** is the second most common (20%). Fundus examination is therefore mandatory in all cases of childhood strabismus.



Fig. 11.54
Left leukocoria due to advanced retinoblastoma (Courtesy of C. Barry)

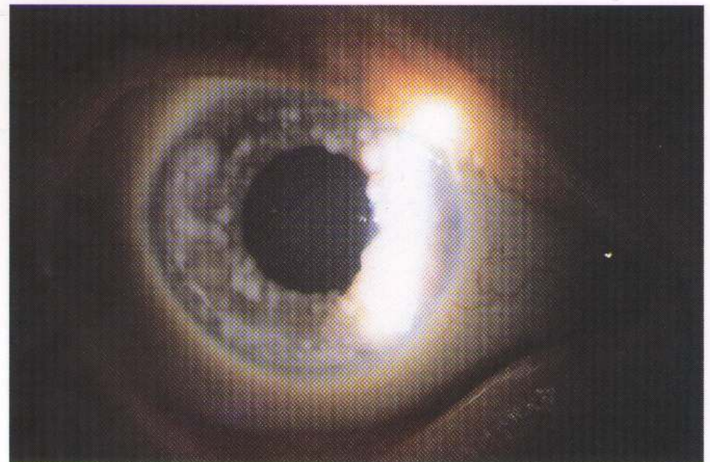


Fig. 11.55
Multiple iris nodules due to invasion by retinoblastoma

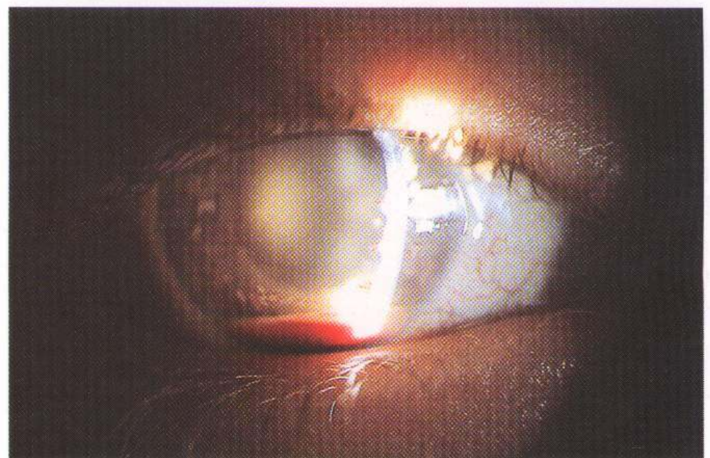


Fig. 11.56
Pseudo-hypopyon due to anterior segment invasion by retinoblastoma

3. **Secondary glaucoma**, sometimes associated with buphthalmos, is uncommon.
4. **Unilateral iris invasion** in older children (average age 6 years) may manifest as multifocal nodules (Fig. 11.55), resembling granulomatous inflammation, or pseudo-hypopyon (masquerade syndrome) (Fig. 11.56). It is therefore important to consider retinoblastoma in the differential diagnosis of unusual chronic uveitis in children.
5. **Orbital inflammation** mimicking orbital or preseptal cellulitis may occur with necrotic tumours (Fig. 11.57). It does not necessarily imply extraocular extension and the exact mechanism is not known.
6. **Orbital invasion** may occur in neglected cases (Fig. 11.58).
7. **Metastatic disease** involving regional lymph nodes and brain before the detection of ocular involvement is rare.
8. **Raised intracranial pressure** due to 'trilateral retinoblastoma' before the diagnosis of ocular involvement is very rare.
9. **On routine examination** of a patient known to be at risk.



Fig. 11.57
Orbital inflammation associated with retinoblastoma



Fig. 11.58
Orbital invasion by neglected retinoblastoma

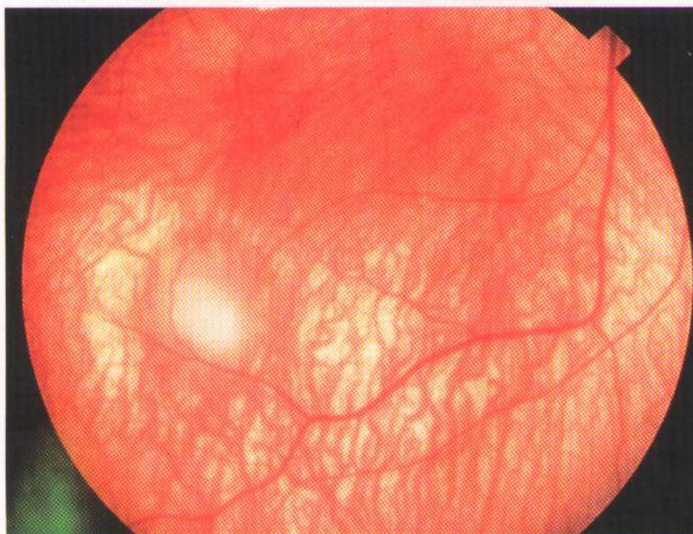


Fig. 11.59
Small intraretinal retinoblastoma

Signs

Indirect ophthalmoscopy with scleral indentation must be performed on *both eyes* after full mydriasis. This is because without indentation pre-equatorial tumours may be missed and one eye may harbour multiple tumours. The clinical signs depend on tumour size and growth pattern.

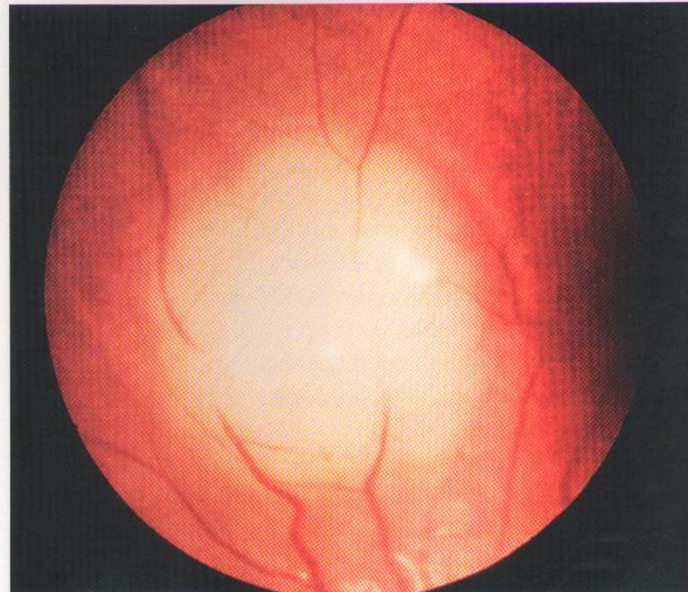


Fig. 11.60
Retinoblastoma

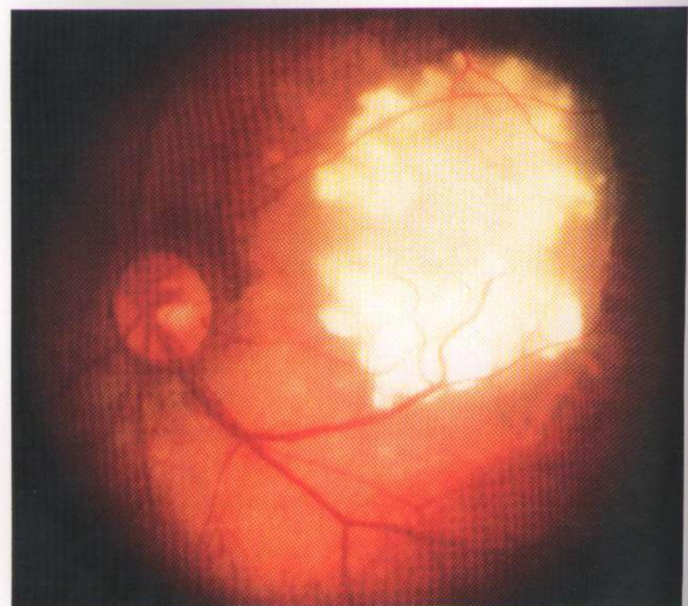


Fig. 11.61
Endophytic retinoblastoma (Courtesy of C. Barry)

1. **An early intraretinal** tumour is a placoid white lesion (Figs 11.59 and 11.60).
2. **An endophytic** tumour grows inwards towards the vitreous, projecting from the retinal surface as a white, cottage cheese-like mass, with surface blood vessels (Fig. 11.61).
3. **An exophytic** tumour grows outwards as a subretinal, multilobulated white mass (Fig. 11.62). It detaches the retina and may be difficult to visualize if the subretinal fluid is deep (Fig. 11.63).



Fig. 11.62
Exophytic retinoblastoma (Courtesy of S. Milewski)

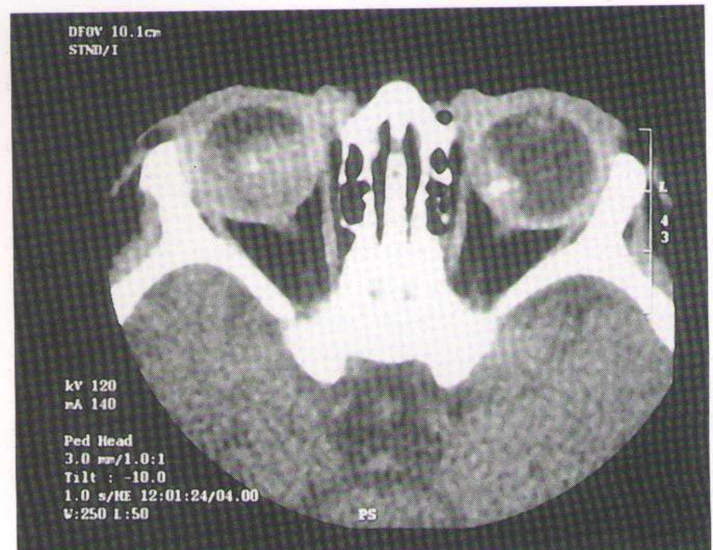


Fig. 11.64
CT scan showing bilateral advanced retinoblastomas (Courtesy of K. Nischal)

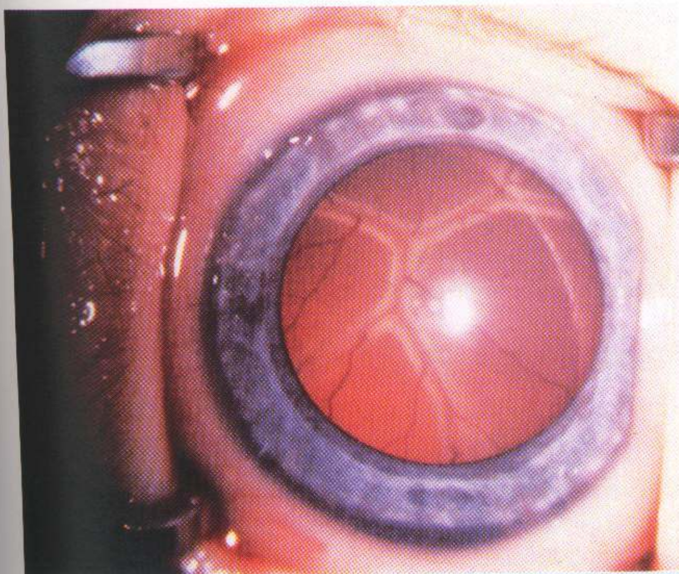


Fig. 11.63
Retinal detachment caused by exophytic retinoblastoma

Special investigations

1. **Ultrasonography** is used mainly to assess tumour size. It also detects calcification within the tumour and is helpful in the diagnosis of simulating lesions such as Coats disease and toxocariasis.
2. **CT** demonstrates gross involvement of the optic nerve, orbital and CNS extension, and the presence of pinealoblastoma and calcification (Fig. 11.64). However, it entails a significant dose of radiation which may be dangerous in patients with germinal mutations.
3. **MRI** cannot detect calcification, but is superior to CT for optic nerve evaluation and detection of a pinealoblastoma, especially when contrast is used. MRI may also be useful to differentiate retinoblastoma from simulating conditions.
4. **Systemic investigations** such as bone marrow aspiration and lumbar puncture are performed only in patients with optic nerve involvement or evidence of extraocular extension.

Treatment

Treatment is related to tumour size, location and associated findings such as retinal detachment, subretinal and vitreous tumour seeds and the state of the fellow eye.

1. **Small tumours**, no more than 4 mm diameter and 2 mm thickness without vitreous or subretinal seeds can be treated with transpupillary thermotherapy laser or cryo-

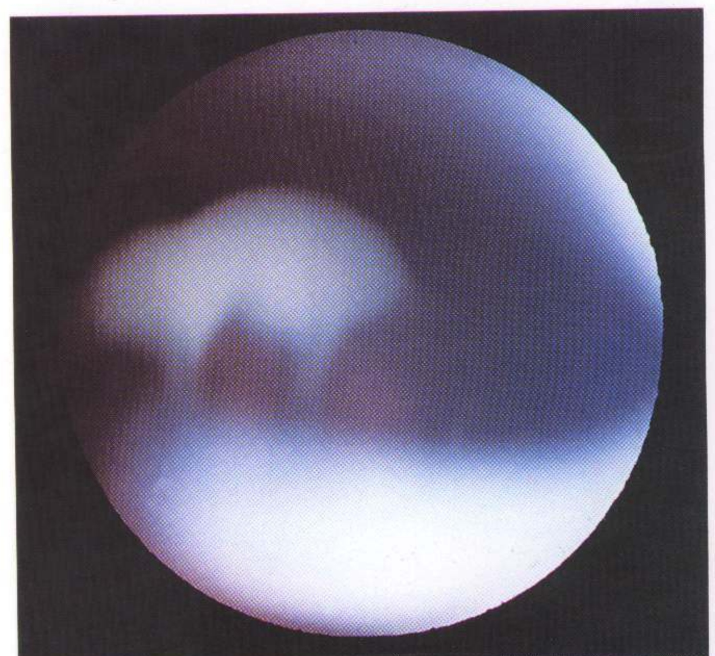


Fig. 11.65
Small peripheral retinoblastoma seen on scleral indentation

therapy. The latter is particularly useful for pre-equatorial tumours which are difficult to reach with laser (Fig. 11.65).

2. **Medium-size tumours**
   a. ***Brachytherapy*** is indicated for tumours of no more than 12 mm diameter and 6 mm thickness, which are unsuitable for thermotherapy or cryotherapy, provided there is no vitreous seeding. Following treatment the tumour regresses leaving a calcific residue (Fig. 11.66).
   b. ***Chemotherapy*** with carboplatin, vincristine and etoposide which may be combined with cyclosporin. The drugs are given intravenously in 3 week cycles over a 4–9 month period depending on disease severity. This may be followed by local treatment with cryotherapy or thermotherapy to consolidate tumour control.

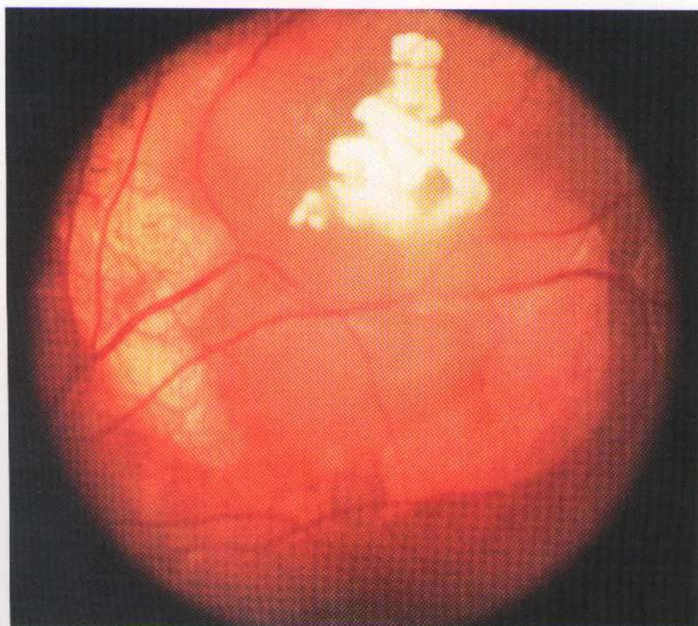


Fig. 11.66
Regressed retinoblastoma with calcific residue

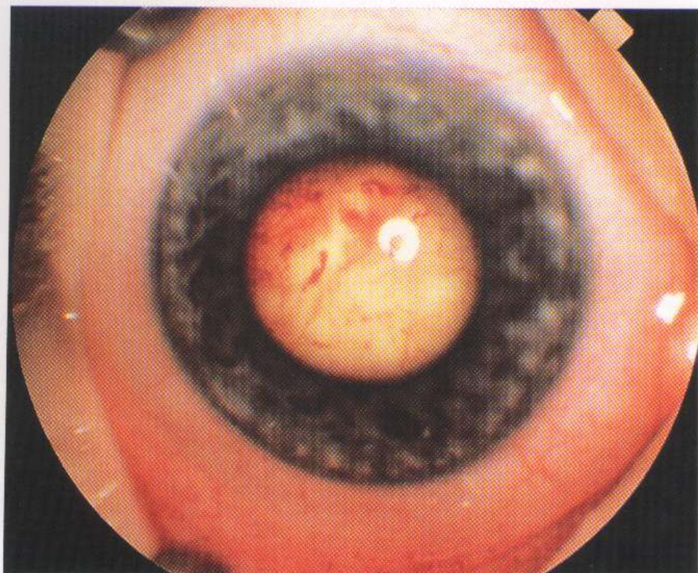


Fig. 11.67
Large retinoblastoma with new surface vessels

   c. ***External beam radiotherapy*** should be avoided, if possible, due to the high risk of complications such as cataract formation, radiation retinopathy and cosmetic deformities. In patients with germinal mutations there is also a risk of inducing a second malignancy such as sarcoma of bone or connective tissue.

3. **Large tumours** (Fig. 11.67)
   a. ***Chemotherapy*** to shrink the tumour (chemoreduction), facilitating subsequent local treatment, thereby avoiding enucleation or external beam radiotherapy. Chemotherapy also will have a beneficial effect if a smaller tumour is present in the fellow eye.
   b. ***Enucleation*** if chemoreduction fails or a normal fellow eye makes aggressive chemotherapy inappropriate. It is also useful for diffuse retinoblastoma because of poor visual prognosis and high risk of recurrence with other therapeutic modalities. Enucleation should be performed with minimal manipulation and it is imperative to obtain a long piece of optic nerve (8–12 mm). There is no contraindication to the insertion of an orbital implant. Unfortunately subsequent shortening of the fornices and retraction of the implant (post-enucleation socket syndrome) may require further surgical intervention.
4. **Extraocular extension** beyond the lamina cribrosa is treated with chemotherapy after enucleation. Extension to the cut end of the optic nerve, or extension through the sclera, is treated with chemotherapy and irradiation of the affected orbit.
5. **Metastatic disease** is treated with high-dose chemotherapy. Patients with malignant cells in the cerebrospinal fluid may require intrathecal methotrexate.

Prognostic factors

The overall mortality rate is 2–5% and is related to the following:

1. **Tumour size and location**. Small posterior tumours do best but there is no significant difference between endophytic and exophytic types.

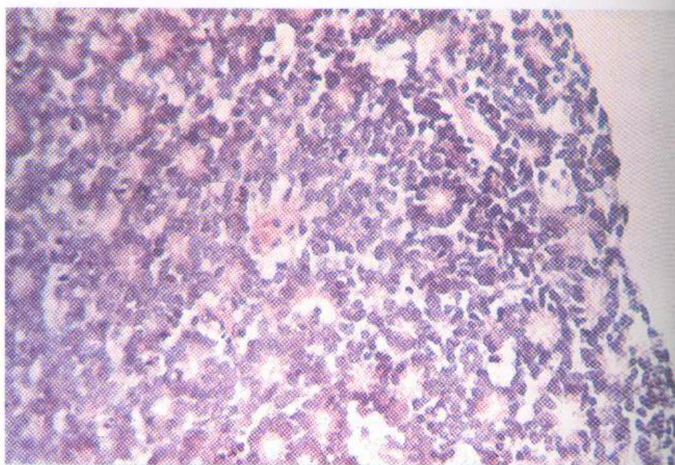
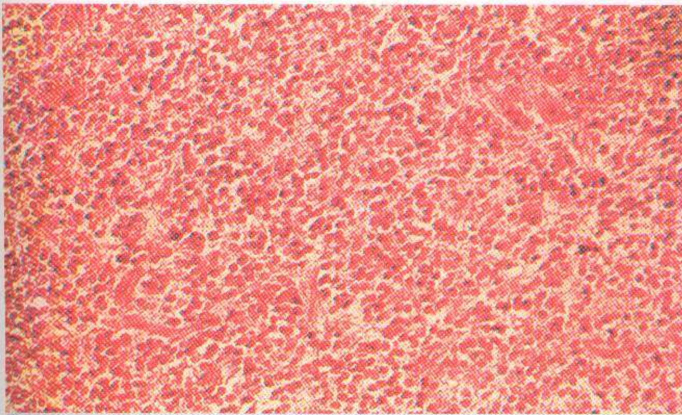


Fig. 11.68
Well-differentiated retinoblastoma with abundant rosettes
(Courtesy of A. Garner)


Fig. 11.69
Highly undifferentiated retinoblastoma (Courtesy of A. Garner)

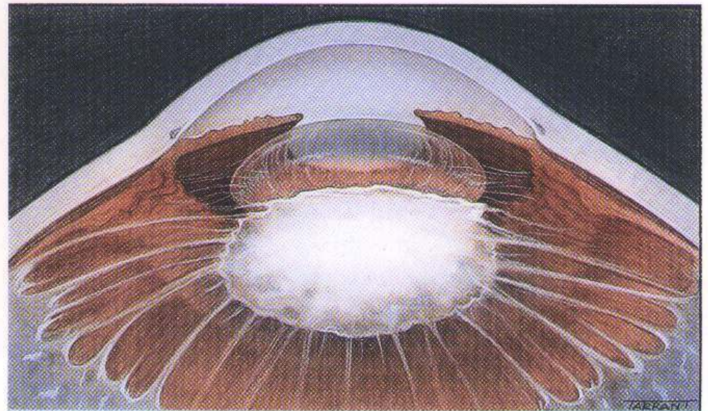
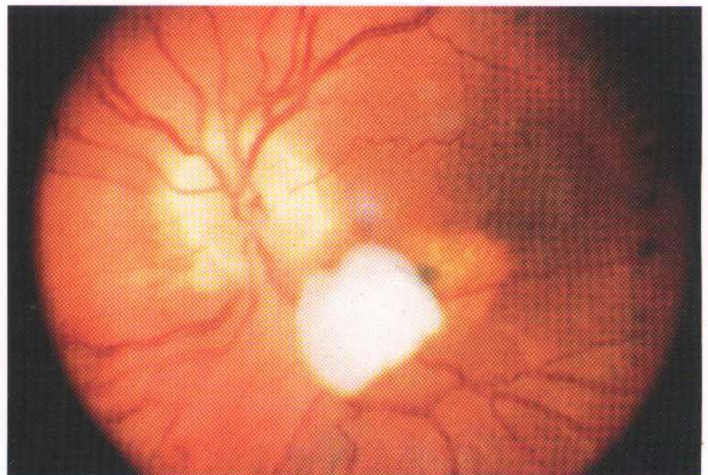

Fig. 11.71
Persistent hyperplastic primary vitreous


Fig. 11.70
Leukocoria associated with persistent hyperplastic primary vitreous


Fig. 11.72
Toxocara granuloma resembling endophytic retinoblastoma

2. **Cellular differentiation**. The mortality rate of patients whose tumours have abundant rosettes (Fig. 11.68) is much less than in those with highly undifferentiated tumours (Fig. 11.69).
3. **Optic nerve involvement** beyond the point of surgical transection is associated with high mortality.
4. **Invasion** of the choroid or vortex veins facilitates haematogenous spread and is therefore of adverse prognostic significance.
5. **Extrascleral spread** carries a grave prognosis.

Differential diagnosis

1. **Persistent hyperplastic primary vitreous** is an important cause of congenital leukocoria (Fig. 11.70). It typically occurs in a microphthalmic eye and is almost always unilateral. It is characterized by a retrolental mass into which elongated ciliary processes are inserted (Fig. 11.71). With time the mass contracts and pulls the ciliary processes centrally so that they become visible through the pupil. An associated dehiscence involving the posterior capsule may lead to subsequent cataract formation.
2. **Coats disease** is almost always unilateral, more common in boys and tends to present later than retinoblastoma. It is characterized by telangiectatic retinal blood vessels, extensive intra- and subretinal yellow exudation and exudative retinal detachment (*see* Chapter 14).
3. **Retinopathy of prematurity**, if advanced, may cause retinal detachment and leukocoria. Diagnosis is usually straightforward because of the history of prematurity and low birth weight (*see* Chapter 14).
4. **Toxocariasis** (*see* Chapter 10).
   a. ***Chronic toxocara endophthalmitis*** may cause a cyclitic membrane and a white pupil.
   b. *Toxocara granuloma* at the posterior pole may resemble an endophytic retinoblastoma (Fig. 11.72).
5. **Intermediate uveitis** may mimic the diffuse infiltrating type of retinoblastoma seen in older children (*see* Chapter 10).
6. **Retinal dysplasia** is characterized by a congenital pink or white retrolental membrane in a microphthalmic eye, with a shallow anterior chamber and elongated ciliary processes. Unilateral cases are usually not associated with systemic abnormalities. Patients with bilateral involvement may



Fig. 11.73
Vesiculobullous dermatitis in incontinentia pigmenti

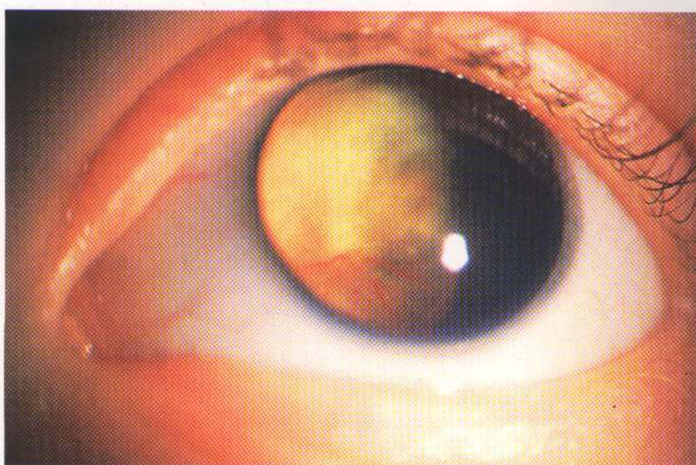


Fig. 11.74
Cicatricial retinal detachment in incontinentia pigmenti

have Norrie disease or Warburg, Patau and Edward syndromes.

7. **Incontinentia pigmenti** (Bloch–Sulzberger syndrome) is a rare X-linked dominant disorder affecting girls. It is characterized by vesiculobullous dermatitis (Fig. 11.73) on the trunk and extremities. Malformations of teeth, hair, nails, bones and central nervous system may also be present. About one-third of children develop cicatricial retinal detachment which may cause leukocoria in the first year of life (Fig. 11.74).
8. **Retinocytoma** (retinoma) is thought to be a benign variant of retinoblastoma. It is characterized by calcified mass associated with RPE alteration and chorioretinal atrophy (Fig. 11.75). The appearance is remarkably similar to that of a retinoblastoma following irradiation (*see* Fig. 11.66).
9. **Retinal astrocytoma** (*see* below).

Retinal astrocytoma

Astrocytoma of the retina or optic nerve head is a rare, benign, non-vision-threatening tumour. It may occur in

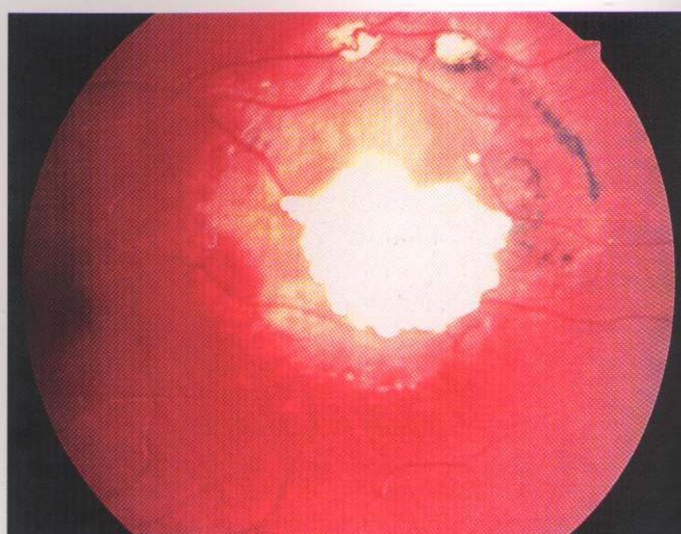


Fig. 11.75
Retinocytoma (Courtesy of K. Nischal)

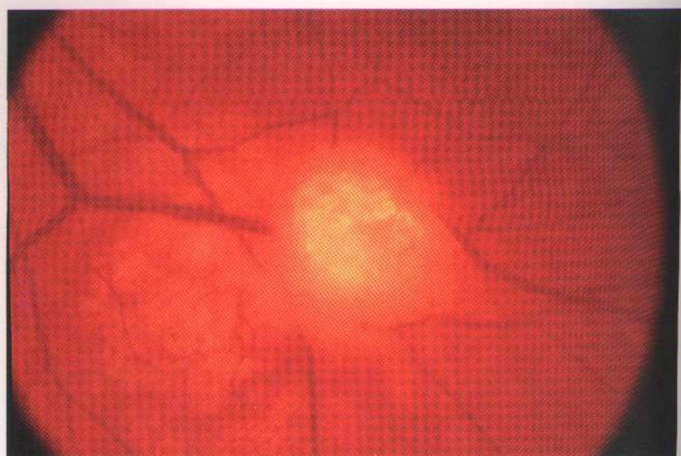


Fig. 11.76
Nodular retinal astrocytoma

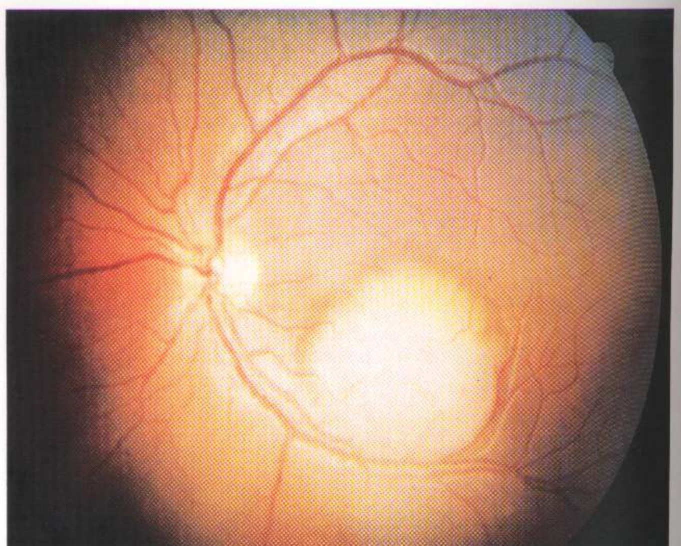


Fig. 11.77
Flat retinal astrocytoma (Courtesy of C. Barry)

isolation but is most frequently seen in patients with tuberous sclerosis (*see* Chapter 20). About 50% of patients with tuberous sclerosis have fundus astrocytomas, which may be multiple and are bilateral in about 15% of cases.

Signs

- A semi-translucent nodule (Fig. 11.76) or a white, relatively flat, well-circumscribed plaque (Fig. 11.77).
- Later, the tumour becomes more solid and white and may, on cursory examination, resemble retinoblastoma (Fig. 11.78).
- Multiple areas of calcification within a long-standing tumour may give rise to a fossilized, mulberry-like appearance (Fig. 11.79).

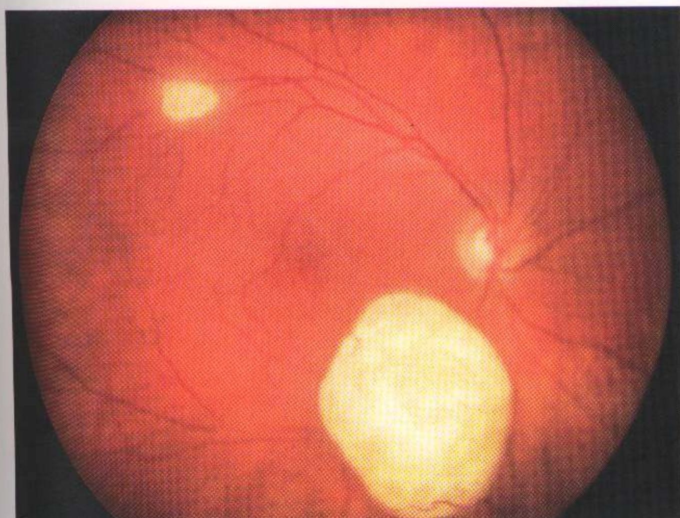


Fig. 11.78
Long-standing retinal astrocytomas

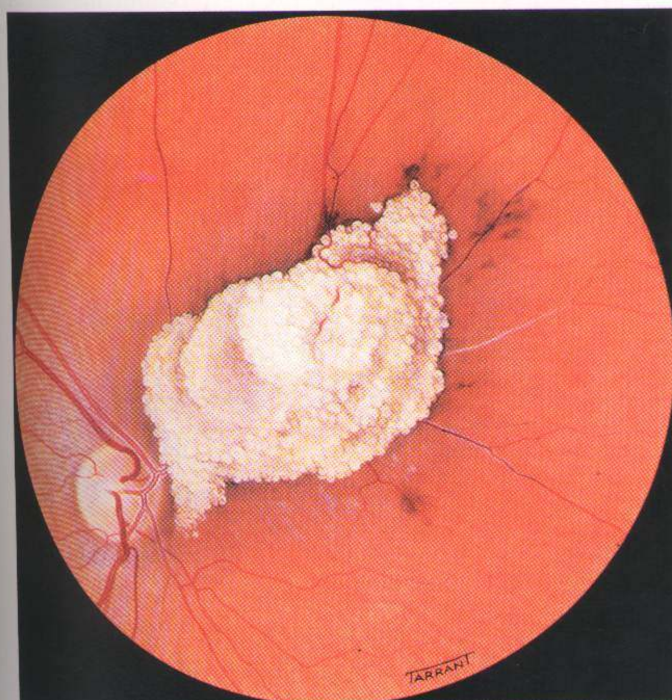


Fig. 11.79
Calcified mulberry-like retinal astrocytoma

Retinal capillary haemangioma

Capillary haemangioma of the retina or optic nerve head is a rare, sight-threatening vascular hamartoma which may occur in isolation (von Hippel disease). However about 50% of patients with solitary capillary haemangiomas and virtually all patients with multiple lesions have systemic disease. The combination of systemic and ocular lesions is referred to as the von Hippel–Lindau syndrome (V-H-L) (*see* Chapter 20). The prevalence of retinal capillary haemangiomas in patients with V-H-L is approximately 60%.

Endophytic

1. **Presentation** is in the second to third decades with unilateral or bilateral ocular involvement on screening, or with visual impairment.
2. **Signs** (in chronological order)
   - A tiny red lesion located within the capillary bed between an arteriole and a venule (Fig. 11.80).

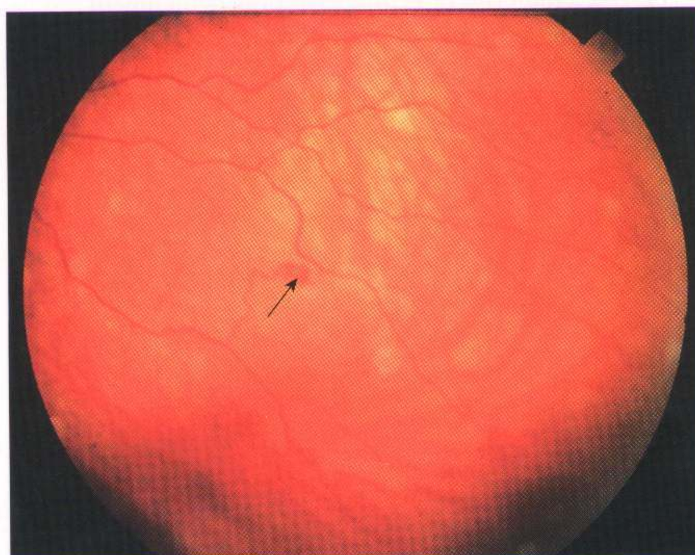


Fig. 11.80
Early retinal capillary haemangioma

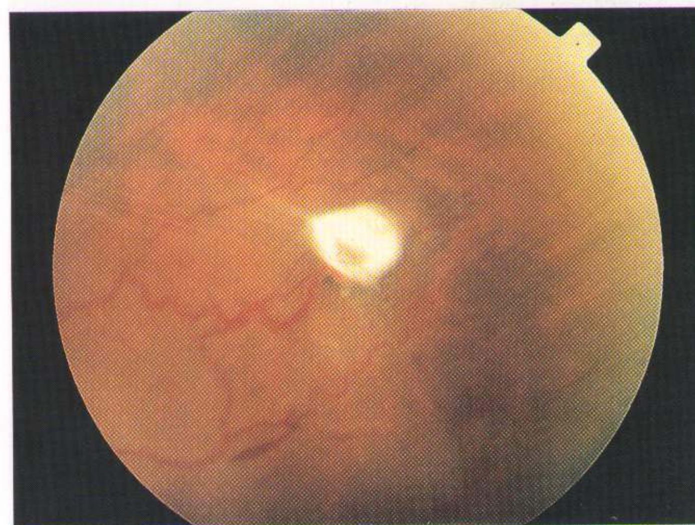


Fig. 11.81
Retinal capillary haemangioma (Courtesy of S. Milewski)

- A small, well-defined nodule (Fig. 11.81).
- A round orange-red mass associated with dilatation and tortuosity of the supplying artery and draining vein due to arteriovenous shunting so that both vessels appear similar (Fig. 11.82).

3. **FA** shows early hyperfluorescence (Fig. 11.83a) and late leakage (Fig. 11.83b).
4. **Complications** include hard exudate formation in the area surrounding the tumour and/or at the macula (*see* Fig. 11.85), macular oedema, epiretinal membrane formation, retinal detachment which may be tractional or exudative, and vitreous haemorrhage.

Other types

1. **Exophytic haemangioma** is less common, arises from the outer retina in the juxtapapillary region and presents with visual loss. It is characterized by a sessile, ill-defined lesion with dilated blood vessels, which may be associated with retinal oedema and haemorrhage (Fig. 11.84). It carries a high risk of exudative retinal detachment.
2. **Optic nerve head haemangioma** (Fig. 11.85)

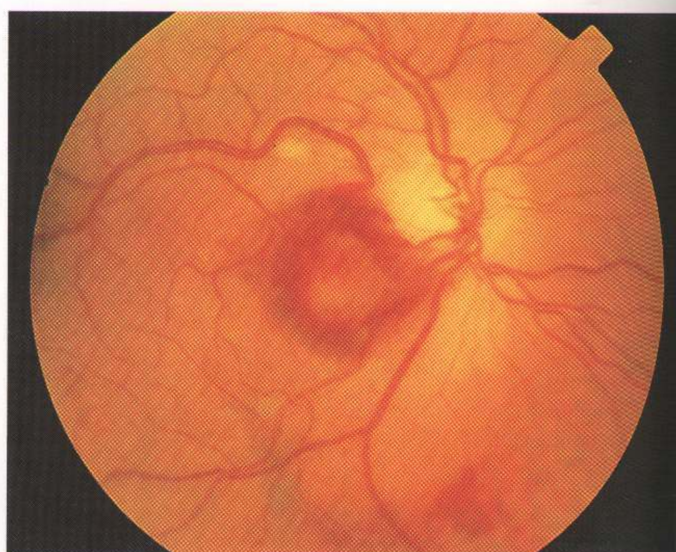


Fig. 11.84
Exophytic retinal capillary haemangioma (Courtesy of S. Milewski)

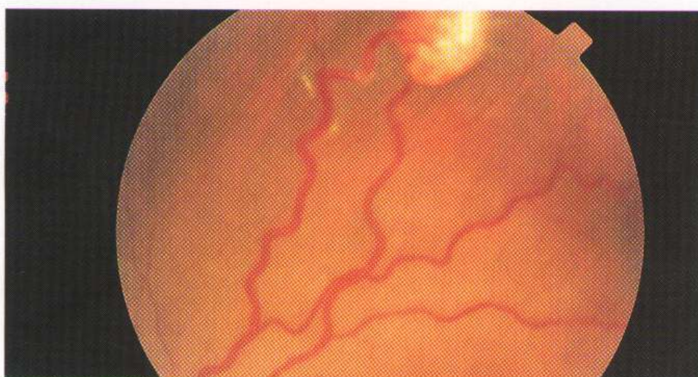
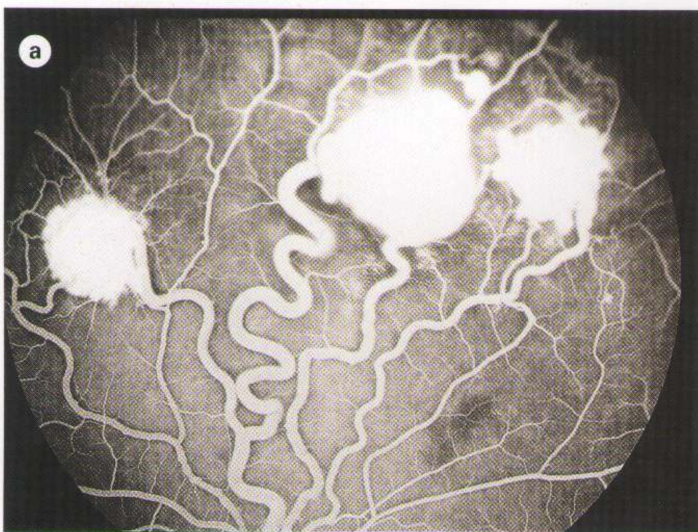


Fig. 11.82
Vascular dilatation and tortuosity associated with a retinal capillary haemangioma

Treatment

1. **Argon laser photocoagulation** for small peripheral lesions (Fig. 11.86a). Following successful treatment the calibre of the feeding blood vessels returns to normal (Fig. 11.86b).
2. **Cryotherapy** for larger peripheral lesions or those with exudative retinal detachment. Vigorous treatment of a large lesion may cause a temporary but extensive exudative retinal detachment.
3. **Brachytherapy** for lesions one to two disc diameters in size.
4. **Vitreoretinal surgery** may be required for non-absorbing vitreous haemorrhage, epiretinal fibrosis or tractional retinal detachment. If appropriate, the tumour may be destroyed by endolaser photocoagulation.



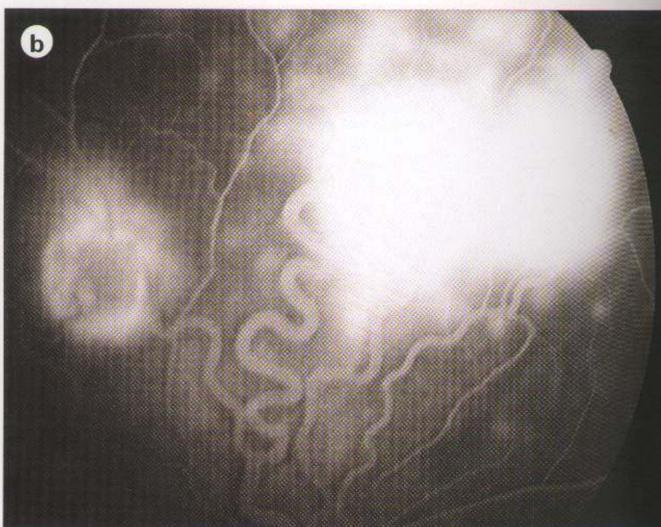


Fig. 11.83
(**a**) Early FA of multiple retinal capillary haemangiomas showing hyperfluorescence due to filling; (**b**) late phase showing leakage (Courtesy of S. Milewski)

Screening

Because it is impossible to predict which patients with retinal angiomas will also harbour systemic lesions, the ophthalmologist must refer all such patients for systemic and neurological evaluation. Relatives should also be screened because of the dominant inheritance pattern of the disease. Apart from physical examination, the following screening protocol should be regularly performed in patients with established V-H-L and relatives at risk:

1. **Annual screening**
   - Eye examination, physical examination and blood pressure measurement.
   - Renal ultrasonography from age 16 years.
   - Twenty-four hour urine collection for estimation of vanillyl mandelic acid and catecholamine levels from age 10 years to detect phaeochromocytoma.
2. **Screening every 3 years** involves abdominal and brain MRI scans from age 15 years.

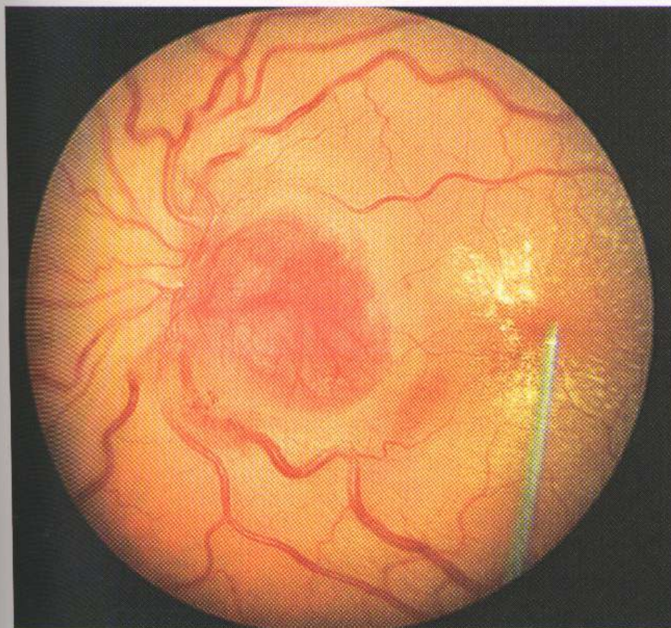


Fig. 11.85
Capillary haemangioma of the optic nerve head with macular exudates (Courtesy of K. Nischal)

Retinal cavernous haemangioma

Cavernous haemangioma of the retina or optic nerve head is a rare, congenital, unilateral, vascular hamartoma. A minority of patients have similar lesions of the skin and CNS. The combination is inherited in autosomal dominant fashion and is referred to as *neuro-oculocutaneous phacomatosis* or alternatively as *cavernoma multiplex*.

1. **Presentation** is in the second to third decades with vitreous haemorrhage, or more often as a chance finding.
2. **Signs** vary from a collection of aneurysms (Fig. 11.87) to an elaborate complex of vascular anomalies on the retina (Fig. 11.88) or optic nerve head (Fig. 11.89) which may rarely bleed (Fig. 11.90). Because of sluggish flow of blood, the red cells may sediment and separate from plasma, giving rise to 'menisci', or fluid levels within the lesion.
3. **Treatment** is generally not required although vitrectomy may be necessary in the rare event of non-absorbing vitreous haemorrhage.



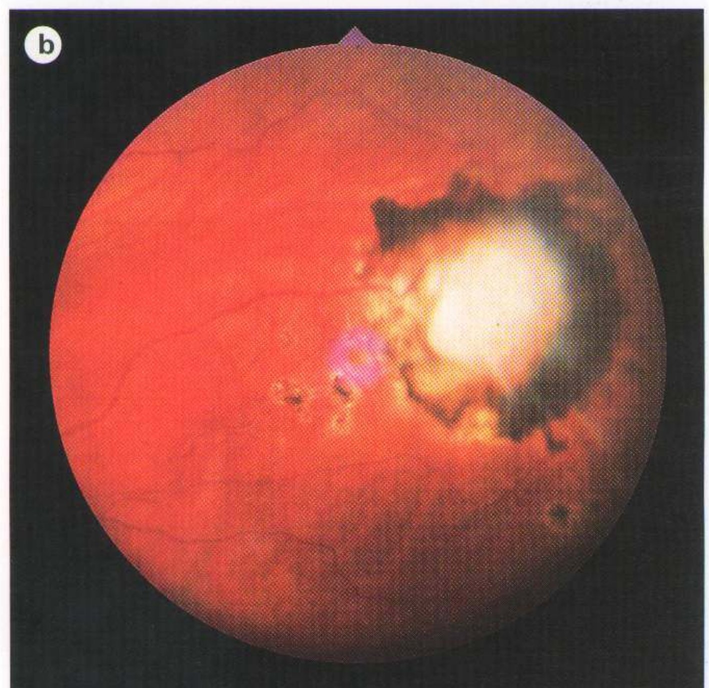


Fig. 11.86
(a) Retinal capillary haemangioma; (**b**) following laser photocoagulation



Fig. 11.87
Small retinal cavernous haemangioma (Courtesy of S. Milewski)

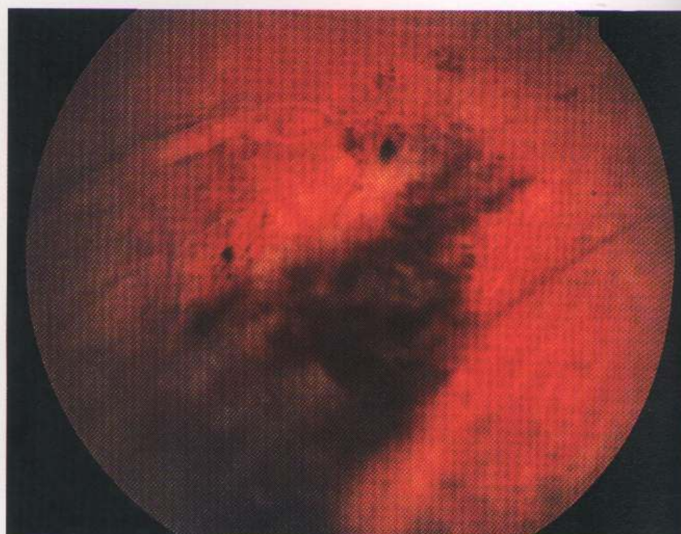


Fig. 11.90
Haemorrhage from retinal cavernous haemangioma

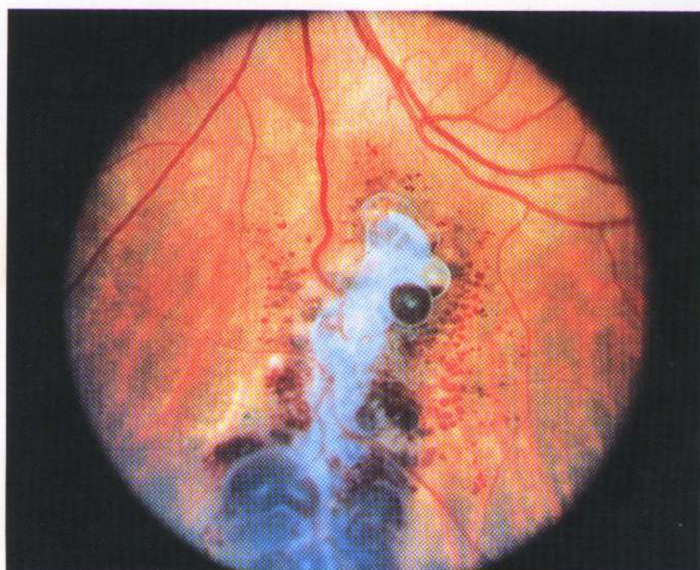


Fig. 11.88
Retinal cavernous haemangioma

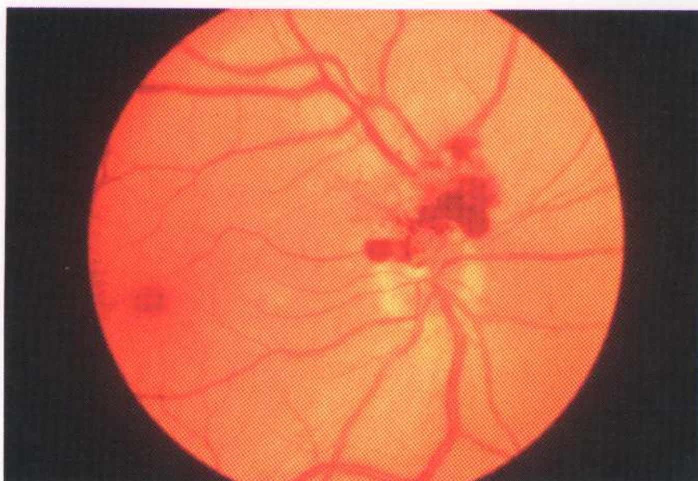


Fig. 11.89
Cavernous haemangioma of the optic nerve head (Courtesy of P. Morse)

Retinal racemose haemangioma

Racemose haemangioma of the retina and optic nerve head is a rare, usually unilateral, congenital arteriovenous malformation involving direct communication between the arteries and veins without an intervening capillary bed. Some patients have similar ipsilateral lesions involving the midbrain, basofrontal region and posterior fossa, an association which is referred to as the *Wyburn–Mason syndrome*. Brain involvement may lead to spontaneous haemorrhage or epilepsy. Occasionally, malformations may involve the maxilla, mandible and orbit. Facial skin lesions have also been reported.

1. **Presentation** may be with visual impairment or more commonly as a chance finding.
2. **Signs**. Enlarged, tortuous blood vessels which are often more numerous than in a normal fundus, with the vein and artery appearing similar (Fig. 11.91a).
3. **FA** shows absence of leakage (Fig. 11.91b–d) but very large lesions may occasionally lead to exudation and haemorrhage.
4. **Treatment** is not required.

Retinal vascular proliferative tumour

Retinal vasoproliferative tumour is a rare gliovascular lesion which occurs mostly in healthy individuals. It can be mistaken for a variety of other entities, most notably retinal angiomas, amelanotic choroidal melanomas and retinal telangiectasia.

1. **Presentation** is in the fifth to sixth decades with blurring of vision due to macular exudation.
2. **Signs**. A solitary, highly vascularized, yellow, retinal or subretinal mass with normal feeding and draining vessels (Fig. 11.92).
3. **Complications** include haemorrhage, exudation, cystoid macular oedema, epiretinal fibrosis and exudative retinal detachment.

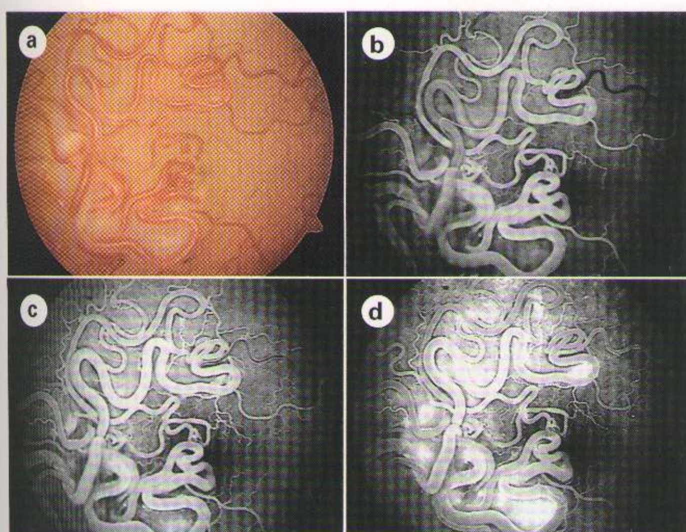


Fig. 11.91
(a) Retinal racemose haemangioma; (b–d) FA showing filling but no leakage (Courtesy of M. Karolczak-Kulesza)

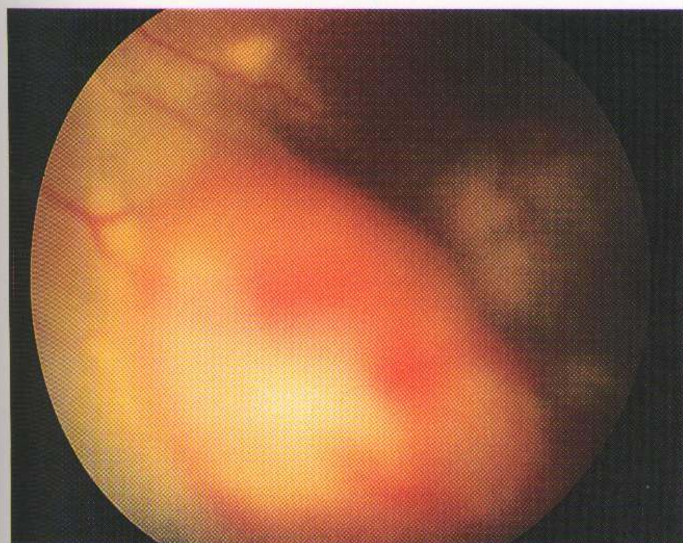


Fig. 11.92
Retinal vascular proliferative tumour (Courtesy of B. Damato)

4. **Treatment** with cryotherapy, laser photocoagulation or brachytherapy may be beneficial but the visual prognosis is guarded.

Tumours of the retinal pigment epithelium

Congenital hypertrophy of the retinal pigment epithelium

Congenital hypertrophy of the retinal pigment epithelium (CHRPE) is a common benign lesion that may be (a) *typical*, which may be solitary or grouped, or (b) *atypical*. It is important to differentiate the two types because the latter has important systemic implications.

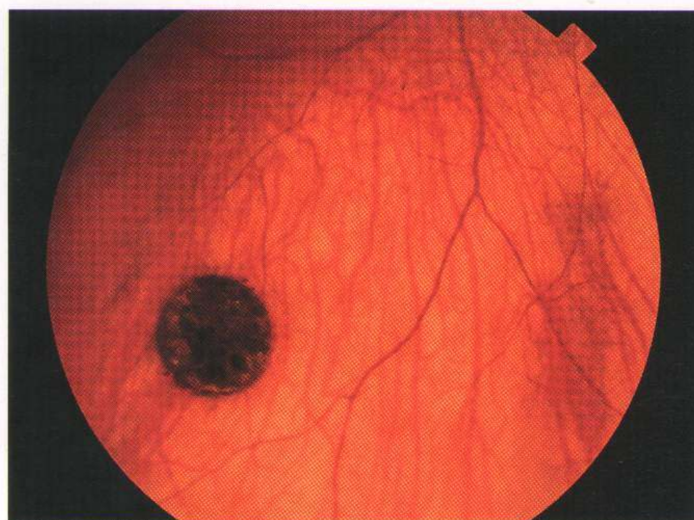


Fig. 11.93
Typical solitary congenital RPE hypertrophy with peripheral depigmentation

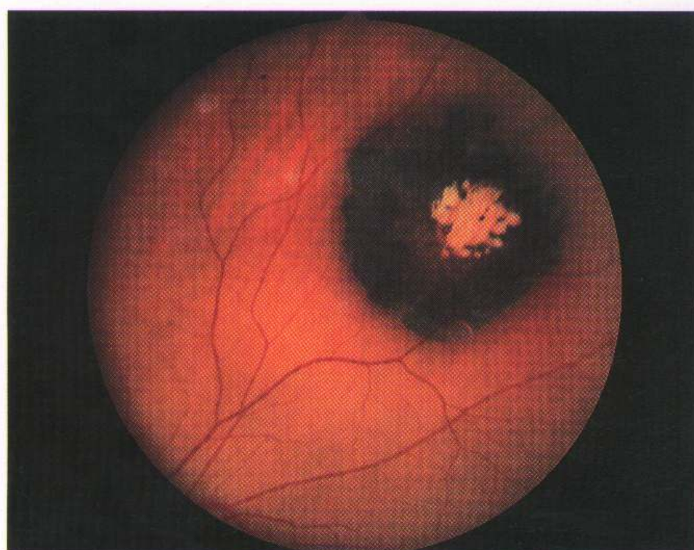


Fig. 11.94
Typical solitary congenital RPE hypertrophy with hypopigmented central lacunae

Typical CHRPE

1. **Solitary**
   - A unilateral, flat, dark-grey or black, well-demarcated, round or oval lesion one to three disc diameters in size frequently with a hypopigmented rim just within the outer margin (Fig. 11.93).
   - Depigmented lacunae which often enlarge or coalesce may be observed, particularly in older patients (Fig. 11.94).
   - Some lesions may become depigmented with only a thin rim of residual pigment remaining at the margin (Fig. 11.95).
2. **Grouped**
   - Usually unilateral, variably sized, sharply circumscribed, round, oval dark-grey or black lesions, often organized in a pattern simulating animal footprints (bear-track pigmentation) (Fig. 11.96).

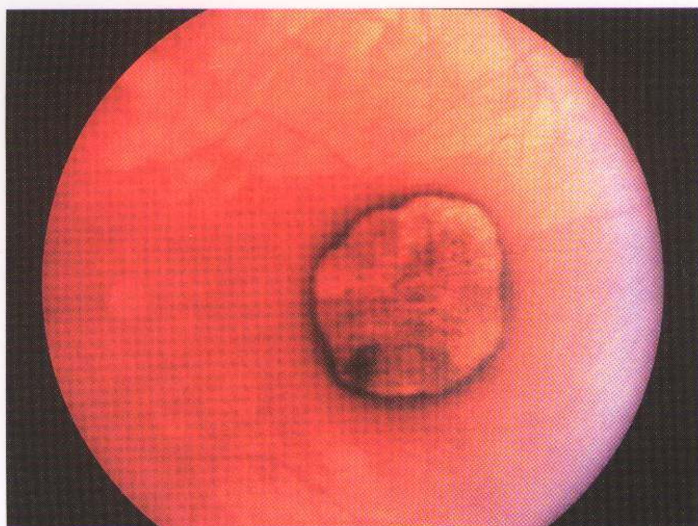


Fig. 11.95
Typical solitary congenital RPE hypertrophy which is nearly completely hypopigmented apart from a peripheral ring

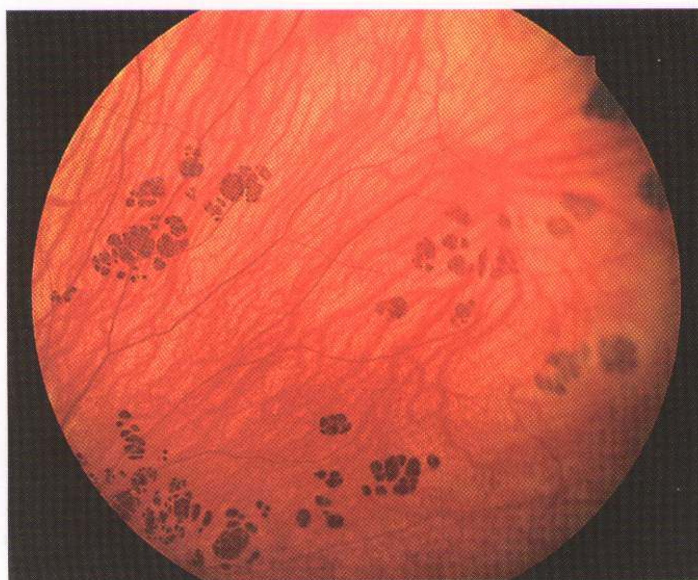


Fig. 11.96
Typical grouped congenital RPE hypertrophy

- The lesions are often confined to one sector or quadrant of the fundus, with the smaller spots usually located more centrally.

Atypical CHRPE

1. Signs

- Multiple, bilateral, widely separated, frequently oval or spindle-shaped lesions of variable size associated with hypopigmentation at one margin (Fig. 11.97).
- The lesions have a haphazard distribution and may be pigmented, depigmented or heterogenous.

2. Systemic implications

a. ***Familial adenomatous polyposis*** (FAP) is a dominantly inherited condition characterized by adenomatous polyps throughout the rectum and colon which usually start to develop in adolescence (Fig. 11.98). If untreated, virtually all patients with FAP develop carcinoma of the colorectal region by the age of 50 years. From the age of 10 years, persons at risk should undergo regular endoscopic examinations and a prophylactic total colectomy should be performed early in adult life in all affected persons. As a result of the dominant inheritance pattern, intensive survey of family members is imperative. The FAP gene has been identified on 5q21–q22. Thus, molecular genetic analysis may identify carriers of the disease in selected cases. Over 80% of patients with FAP have atypical CHRPE which is present at birth. A positive criterion for FAP is the presence of at least four lesions whatever their size, or at least two lesions one of which must be large. Such fundus lesions in a family member should therefore arouse suspicion of FAP.

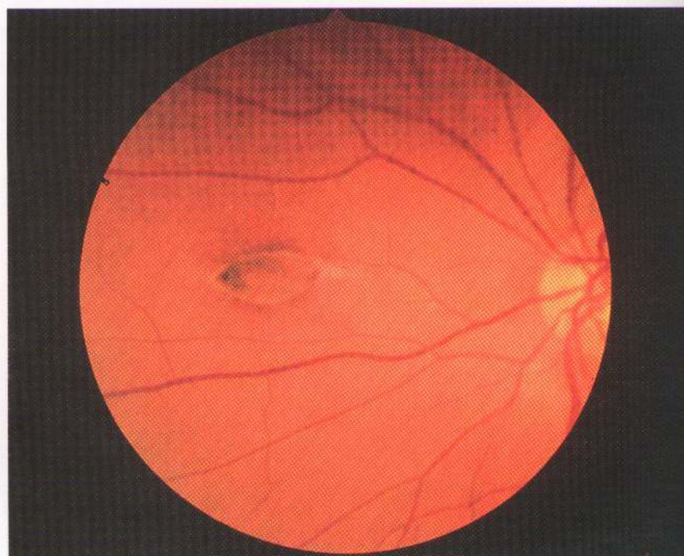


Fig. 11.97
Atypical congenital RPE hypertrophy (Courtesy of B. Jay)

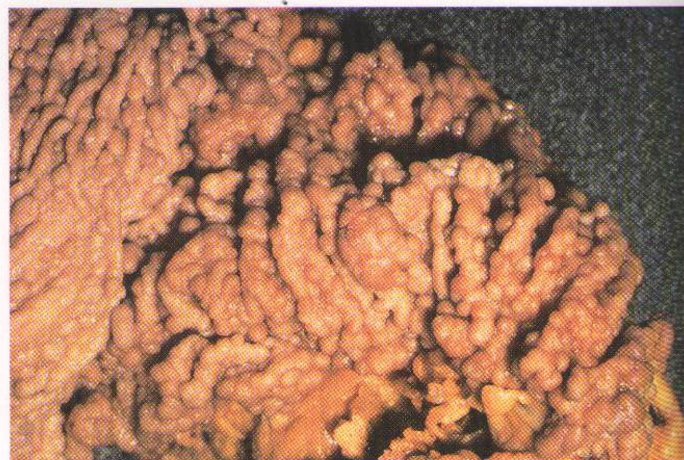


Fig. 11.98
Intestinal adenomatous polyposis

b. ***Gardner syndrome*** is characterized by FAP, osteomas of the skull and mandible and cutaneous soft tissue tumours such as epidermoid cysts, lipomas and fibromas.

c. ***Turcot syndrome*** is characterized by FAP and tumours of the CNS, particularly medulloblastoma and glioma.

Combined hamartoma of the retinal pigment epithelium and retina

Combined hamartoma of the RPE and retina is a rare, usually unilateral malformation which may be juxtapapillary or peripheral. It predominantly affects males and is found with increased frequency in patients with neurofibromatosis-2.

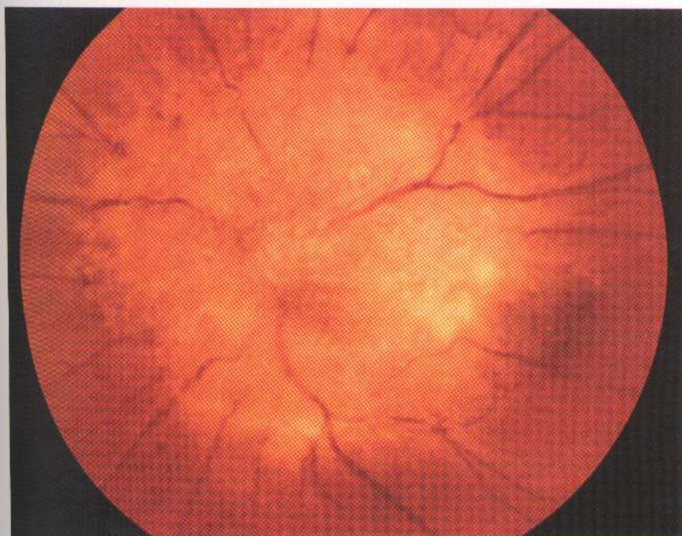


Fig. 11.99
Juxtapapillary combined hamartoma of the RPE and retina

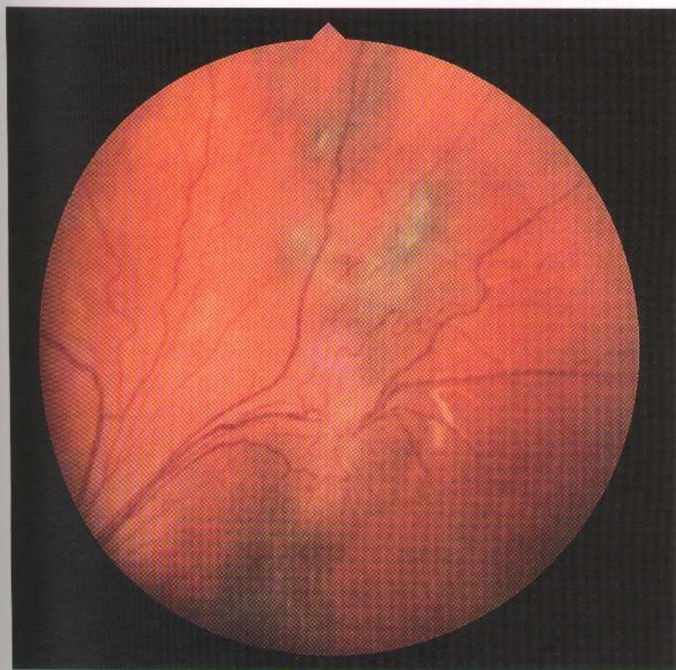


Fig. 11.100
Distortion of the retina by a combined hamartoma of the RPE and retina

1. **Juxtapapillary**
   a. ***Presentation*** is in late childhood or early adulthood with blurred vision and metamorphopsia.
   b. ***Signs***. Deep, slightly elevated, greyish-brown pigmentation associated with variable intra- and epiretinal gliosis, a fine network of dilated capillaries and vascular tortuosity (Fig. 11.99).
2. **Peripheral**
   a. ***Presentation*** is in early childhood with strabismus.
   b. ***Signs***. A linear ridge associated with stretched blood vessels.
3. **Complications** include retinal and/or optic nerve head distortion (Fig. 11.100), macular oedema, choroidal neovascularization and, rarely, retinoschisis and retinal detachment.
4. **Treatment** of epiretinal membranes by vitreoretinal surgery may be tried but the visual results are often disappointing.

Hamartoma of the retinal pigment epithelium

This is an uncommon, small, jet-black lesion involving the RPE, often at the macula, which has a tendency to spill onto the surrounding inner retinal surface (Fig. 11.101).

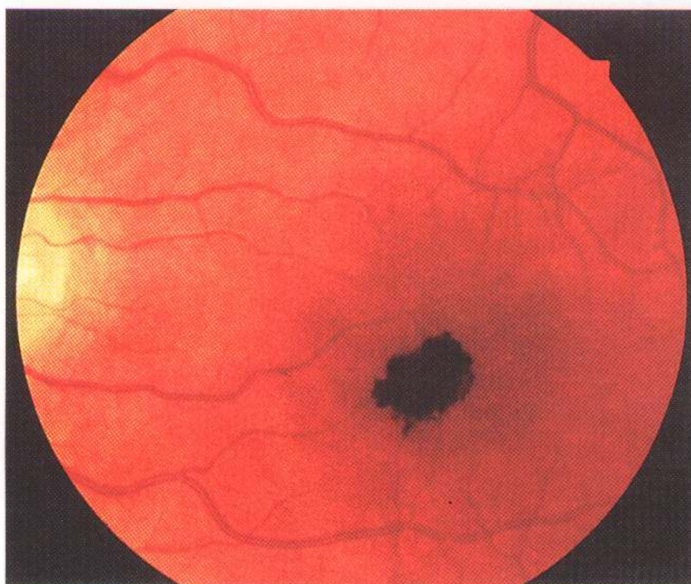


Fig. 11.101
Hamartoma of the RPE

CHAPTER 12

Retinal Detachment

Introduction

Definitions

Retinal detachment

A retinal detachment (RD) is a separation of the sensory retina from the retinal pigment epithelium (RPE) by subretinal fluid (SRF).

1. **Rhegmatogenous RD** (*rhegma*: break) occurs secondarily to a full-thickness defect in the sensory retina, which permits SRF derived from synchitic (liquefied) vitreous gel to gain access to the subretinal space.
2. **Non-rhegmatogenous RD** may be:
   a. *Tractional*, in which the sensory retina is pulled away from the RPE by contracting vitreoretinal membranes; the source of SRF is unknown. Important causes include: proliferative diabetic retinopathy, retinopathy of prematurity, sickle cell retinopathy and penetrating posterior segment trauma.
   b. *Exudative* (serous, secondary), in which SRF derived from the choriocapillaris gains access to the subretinal space through damaged RPE. Important causes include choroidal tumours, exophytic retinoblastoma, Harada disease, posterior scleritis, subretinal neovascularization and severe hypertension.

Vitreoretinal traction

Vitreoretinal traction is a force exerted on the retina by structures originating in the vitreous and may be dynamic or static. The difference between the two is crucial to understanding the pathogenesis of the various types of RD.

1. **Dynamic** traction is induced by rapid eye movements and exerts a centripetal force towards the vitreous cavity. It is relevant to the pathogenesis of retinal tears and rhegmatogenous RD.
2. **Static** traction is independent of ocular movements and plays an important role in the pathogenesis of tractional RD and proliferative vitreoretinopathy. Static traction may be:
   a. *Tangential* (surface) which acts parallel to the surface of the retina as a result of contraction of epiretinal or subretinal membranes.
   b. *Anteroposterior* in which the retina is pulled anteriorly towards the vitreous base.
   c. *Bridging* (trampoline) in which traction occurs between one part of the retina and another, usually along the detached posterior hyaloid surface.

Retinal breaks

Retinal breaks are full-thickness defects of the sensory retina. They can be classified according to (a) *pathogenesis*, (b) *morphology* and (c) *location*.

Pathogenesis

1. **Tears** are caused by dynamic vitreoretinal traction. They have a predilection for the superior fundus (temporal more than nasal).
2. **Holes** are caused by chronic atrophy of the sensory retina and may be round or oval. They have a predilection for the temporal fundus (superior more than inferior) and are less dangerous than tears.

Morphology

Retinal tears may have one of five configurations:

1. **U-tears** (arrowhead tears) (Fig. 12.1a) consist of a flap, the apex of which is pulled anteriorly by the vitreous, the base remaining attached to the retina. The actual tear consists of two anterior extensions (horns), meeting at the apex, which points towards the posterior pole.
2. **Incomplete U-tears** may be linear (Fig. 12.1b), L-shaped (Fig. 12.1c) or J-shaped.
3. **Operculated tears** (Fig. 12.1d) in which the flap is completely avulsed by the detached vitreous gel.
4. **Dialyses** (Fig. 12.1e) are circumferential tears along the ora serrata with vitreous attachment to their posterior margins.

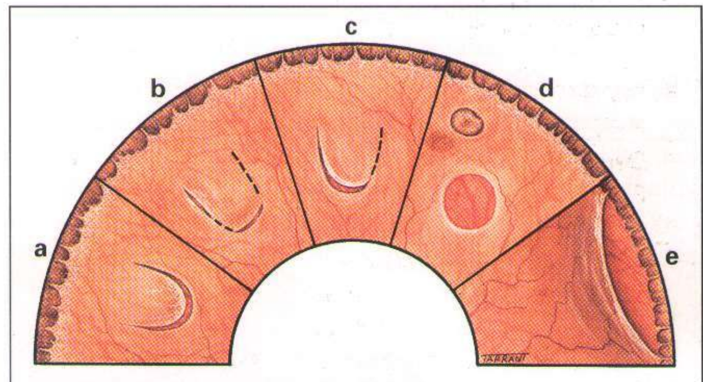


Fig. 12.1
Retinal tears. **(a)** U-shaped; **(b)** linear; **(c)** L-shaped; **(d)** operculated; **(e)** dialysis



Fig. 12.2
Giant retinal tear

5. **Giant tears** (Fig. 12.2) involve 90° or more of the circumference of the globe. They are a variant of U-shaped tears with the vitreous gel attached to the anterior margin of the break. Giant tears are most frequently located in the immediate post-oral retina and less commonly at the equator.

Location

1. **Oral** breaks are located within the vitreous base.
2. **Post-oral** breaks are located between the posterior border of the vitreous base and equator.
3. **Equatorial** breaks are at or near the equator.
4. **Post-equatorial** breaks are behind the equator.
5. **Macular** breaks, invariably holes, are at the macula.

Applied anatomy

Ora serrata

The ora is the junction between the retina and ciliary body (Fig. 12.3). The nasal ora is characterized by teeth-like extensions of retina onto the pars plana (dentate processes) separated by oral bays (Fig. 12.4). In the temporal ora the dentate processes are blunt or absent. Microcystoid degeneration is a normal finding involving the perioral retina characterized by tiny vesicles with indistinct boundaries which make the retina appear thickened and less transparent (Fig. 12.5). At the ora, fusion of the sensory retina with the RPE and choroid limits further forward extension of RD. However, there being no equivalent adhesion between the choroid and sclera, choroidal detachments invariably progress anteriorly to involve the ciliary body (ciliochoroidal

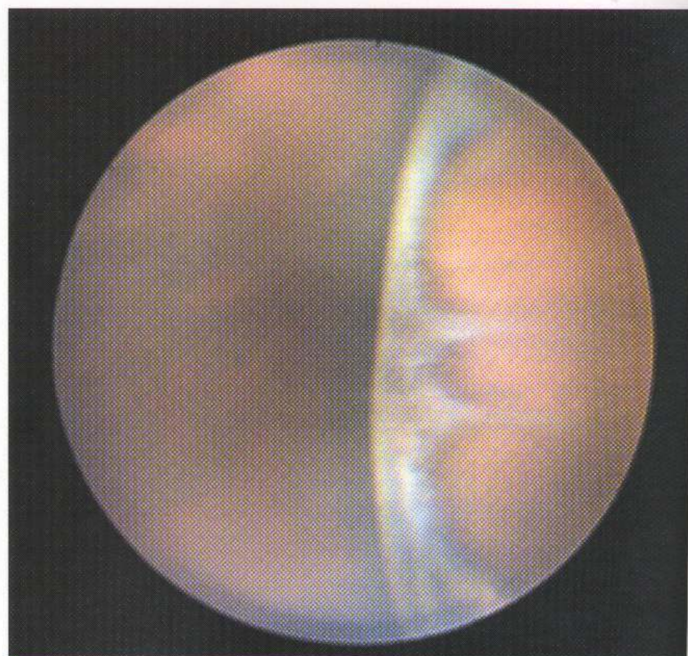


Fig. 12.4
Dentate processes and oral bays (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence California, 1982)

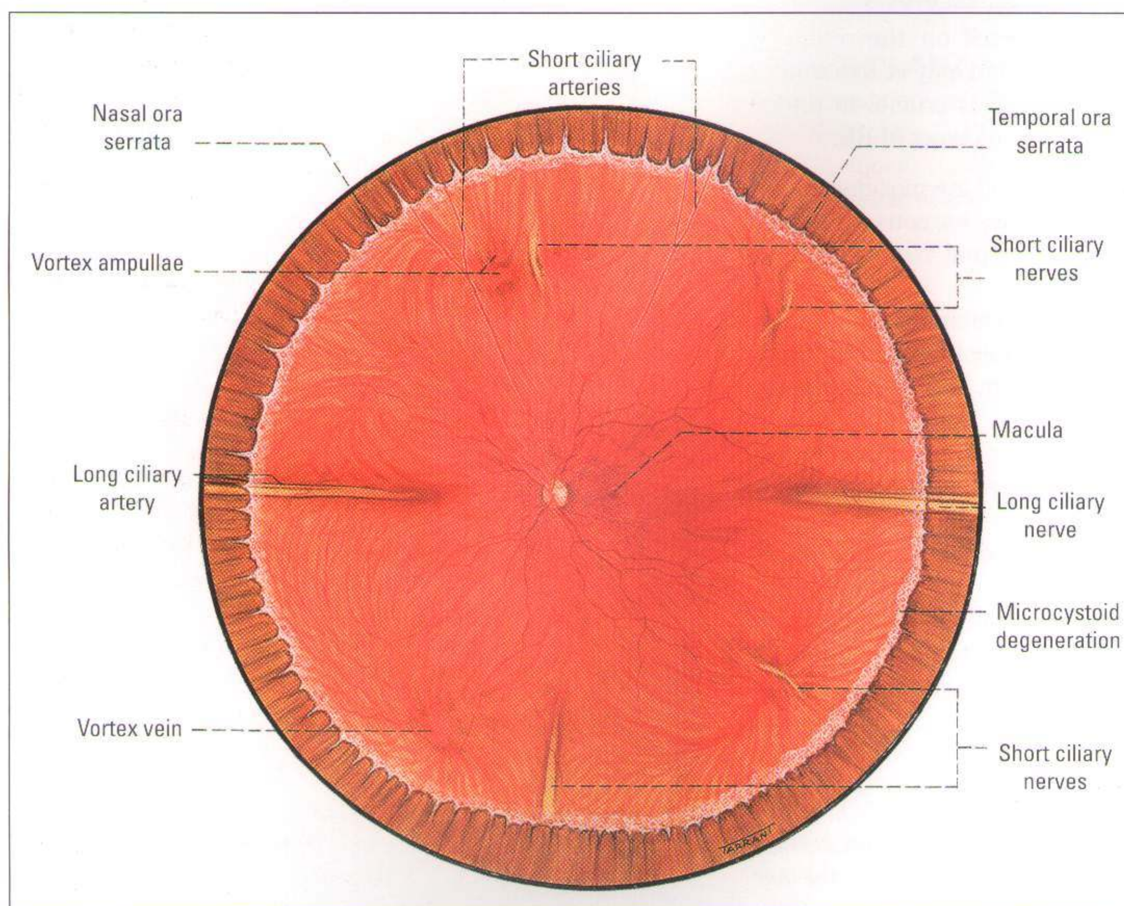


Fig. 12.3
Normal anatomical landmarks

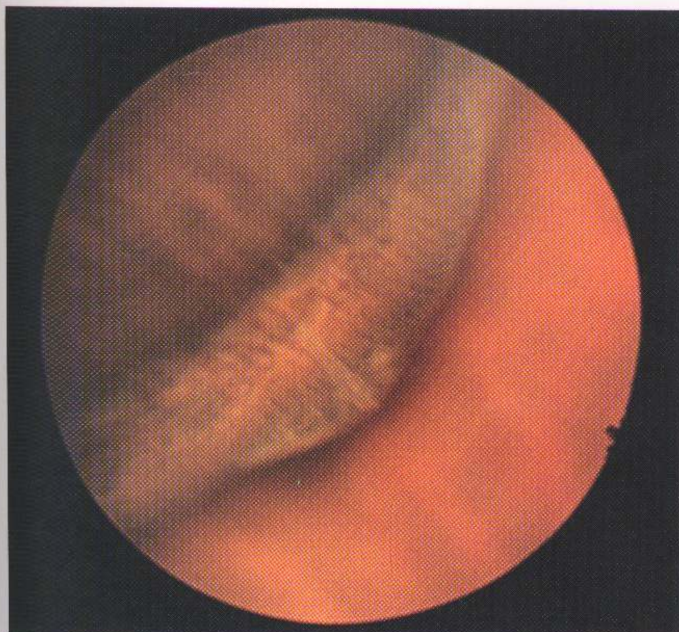


Fig. 12.5
Microcystoid degeneration (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Altas*. Criterion Press, Torrence California, 1982)

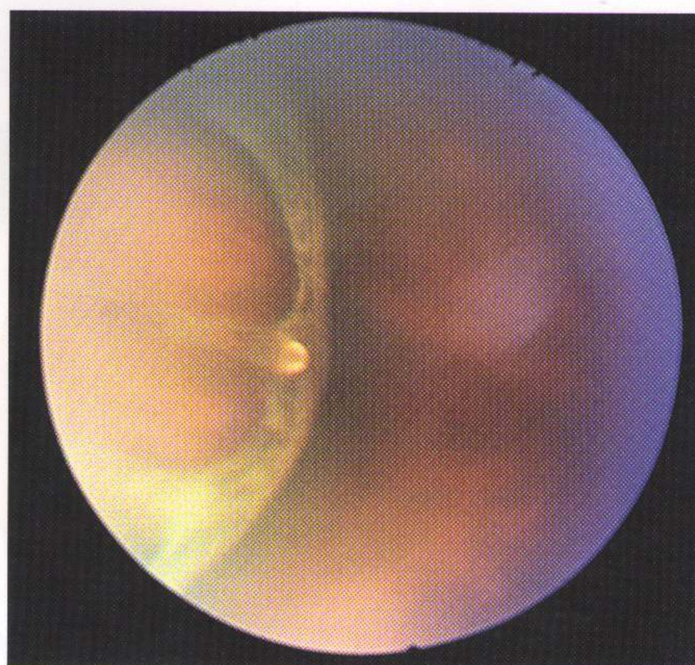


Fig. 12.7
Oral pearl (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence California, 1982)

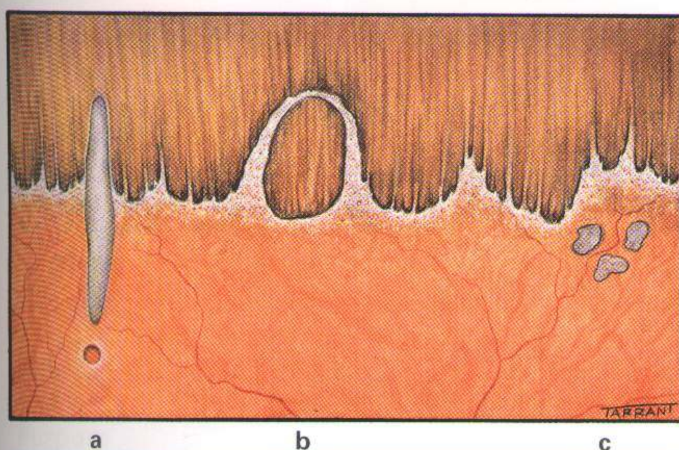


Fig. 12.6
Normal variants of the ora serrata. **(a)** Meridional fold with a small hole at its base; **(b)** enclosed oral bay; **(c)** granular tissue

detachment). The following congenital anomalies may occasionally have clinical significance (Fig. 12.6):

1. **A meridional fold** is a small radial fold of retina in line with a dentate process. It may occasionally exhibit a small retinal hole at its base (Fig. 12.6a).
2. **An enclosed oral bay** is a small island of pars plana surrounded by retina as a result of meeting of two adjacent dentate processes (Fig. 12.6b). It should not be mistaken for a retinal hole because it is located anterior to the ora serrata.
3. **Granular tissue** characterized by multiple, tiny, white opacities within the vitreous base (Fig. 12.6c) can sometimes be mistaken for very small peripheral opercula.
4. **Oral pearls** (Fig. 12.7) are uncommon tiny white lesions.

Vitreous base

This is a 3–4 mm wide zone straddling the ora serrata (Fig. 12.8). The cortical vitreous is strongly adherent at the vitreous base, so that following acute posterior vitreous detachment (PVD) the posterior hyaloid surface remains attached to the posterior border of the vitreous base. Pre-existing perioral retinal holes within the vitreous base therefore do not lead to RD.

Vitreoretinal adhesions

1. **Normal adhesion** between the cortical vitreous and the internal limiting membrane is fairly loose, except at the following sites:
   - At the vitreous base (very strong).
   - Around the optic disc margin (fairly strong).
   - Around the fovea (weak).
   - Around peripheral retinal blood vessels (usually weak).
2. **Abnormal adhesions** may occasionally be associated with retinal tear formation caused by dynamic vitreoretinal traction in eyes with acute PVD. They occur at the following sites:
   - Posterior border of lattice degeneration.
   - Congenital cystic retinal tufts which are white, ovoid, inward projections of sensory retina located in the postoral area (Fig. 12.9).
   - Retinal pigment clumps.
   - Peripheral paravascular condensations.
   - Vitreous base anomalies such as posterior tongue-like extensions and isolated islands.
   - Areas of 'white-without-pressure'.

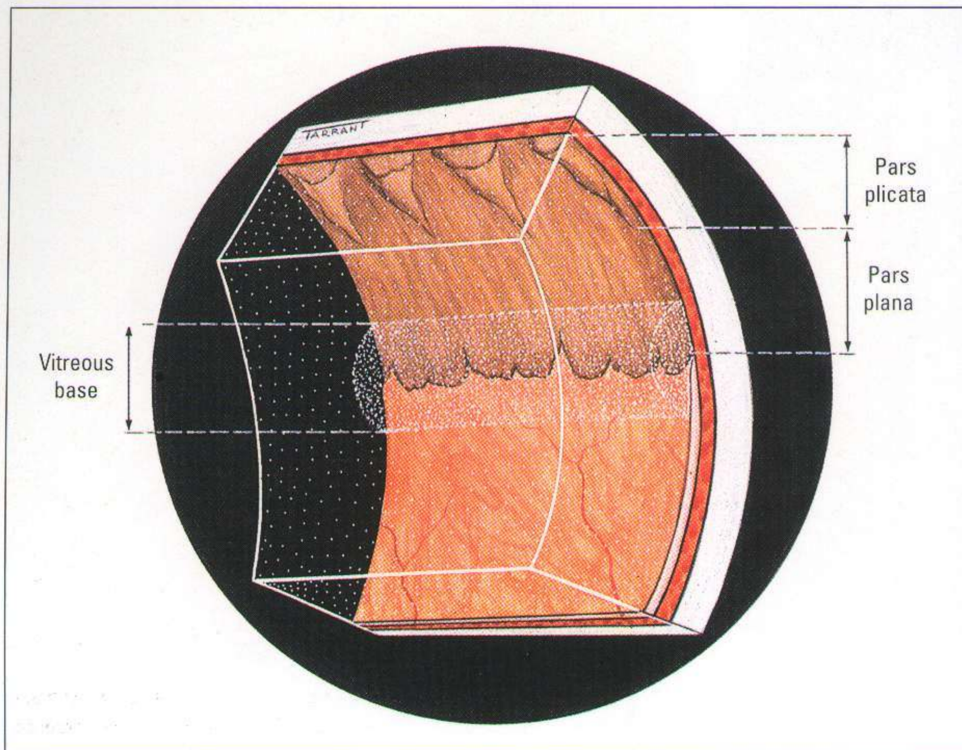


Fig. 12.8
Anatomy of the vitreous base

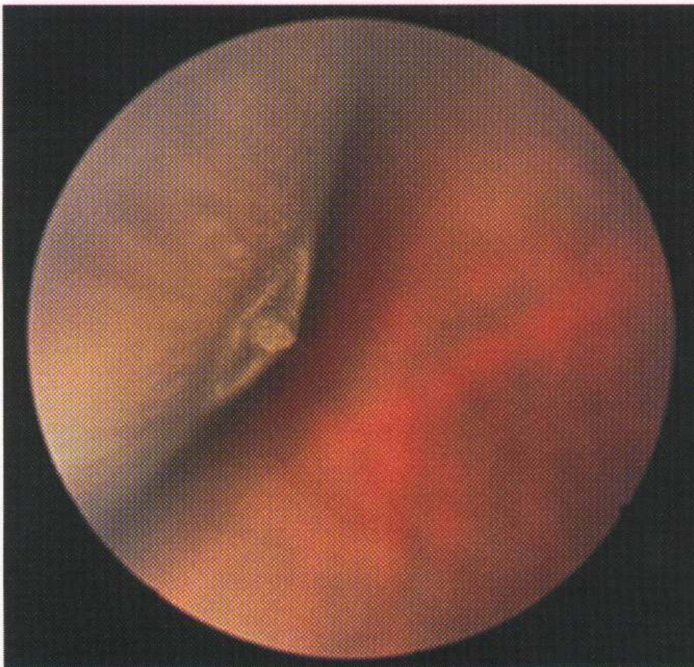


Fig. 12.9
Cystic retinal tuft (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence, California, 1982)

Blood vessels

1. **The long posterior ciliary arteries** (accompanied by corresponding nerves) are recognized as yellow lines that start behind the equator and run anteriorly in the 3 and 9 o'clock meridians (*see* Fig. 12.3). They divide the fundus into upper and lower zones. The arteries run in the suprachoroidal space in line with the horizontal recti.
2. **The short posterior ciliary arteries** are unaccompanied by nerves and may be difficult to identify ophthalmoscopically. The short ciliary nerves appear as peripheral radial yellow lines.
3. **The vortex ampullae** are located just posterior to the equator in the 1, 5, 7 and 11 o'clock meridia. Externally the vortex veins emerge from their scleral canals at variable distances from the equator. They limit posterior extension of a choroidal detachment as they pass through the suprachoroidal space into their scleral canals.

Examination techniques

Indirect ophthalmoscopy

Condensing lenses of various powers and diameters are available (Fig. 12.10). The higher the power, the lower the magnification, the shorter the working distance but the greater the field of view. The technique is as follows:

1. Both pupils should be well dilated.
2. The patient should ideally be supine.
3. The lens is held with the flat surface facing the patient and should be held parallel to the patient's iris plane at all times.
4. The red reflex and then the fundus are located.
5. The tendency to move towards the patient should be avoided if difficulty in visualizing the fundus is encountered.
6. The patient is asked to move the eyes and head into optimal positions for examination.



Fig. 12.10
Condensing lenses used for indirect ophthalmoscopy

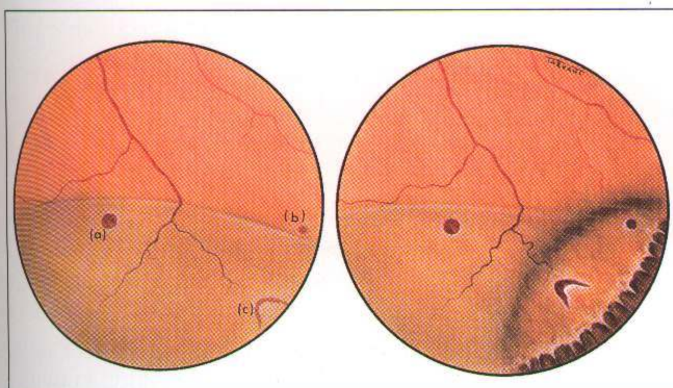


Fig. 12.11
(Left) Retinal breaks in detached retina without scleral indentation; **(right)** with scleral indentation

Scleral indentation

Purpose

Scleral indentation enhances visualization of the peripheral retina anterior to the equator and enables a dynamic fundus

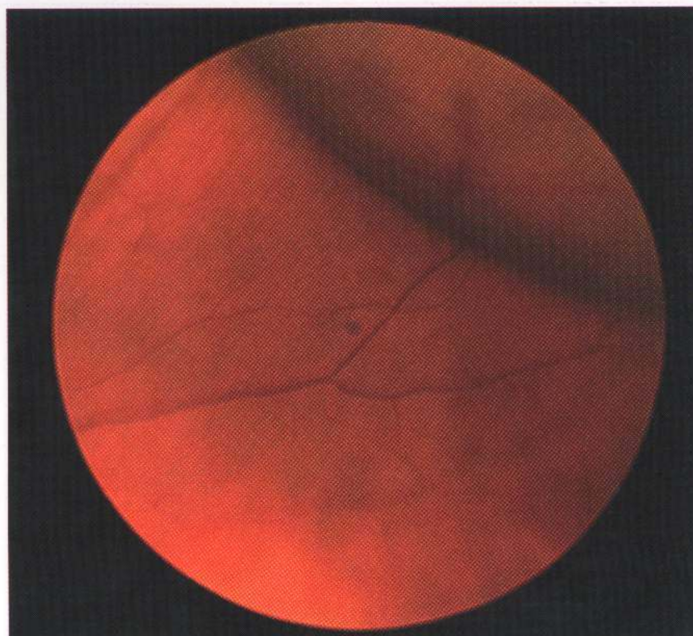
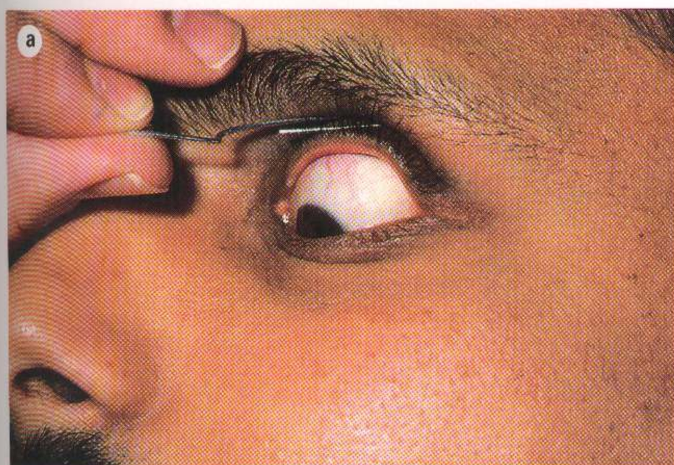


Fig. 12.13
Mound created by scleral indentation (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence California, 1982)

examination to be performed. For example, Fig. 12.11 (left) shows a retinal hole (a) near the equator, visible without scleral indentation because the underlying choroid provides good contrast and the hole appears red. However, a small round hole (b) near the ora serrata or a small U-tear (c) near the posterior border of the vitreous base may be overlooked without scleral indentation. Figure 12.11 (right) shows that with scleral indentation the small hole (b) is seen more easily because the contrast between choroid and sensory retina is enhanced. Indentation also brings the peripheral fundus into view and enables the flap of the small U-tear (c) to be seen in profile.

Technique

1. To view the ora serrata at 12 o'clock, the patient is asked to look down. The scleral indentor is applied to the outside



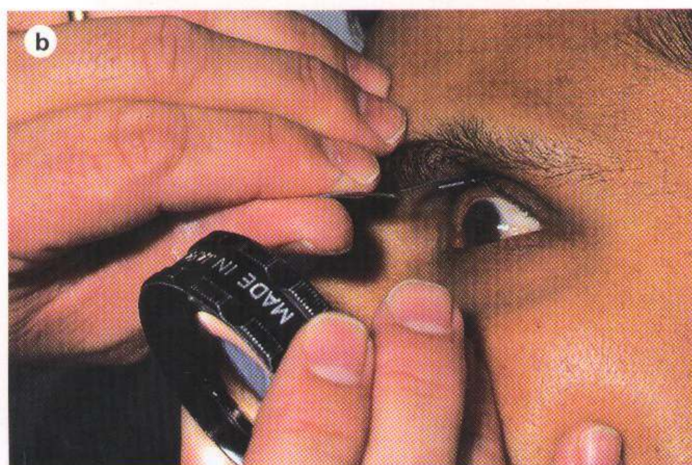


Fig. 12.12
(a) Insertion of an unfolded paper clip; (**b**) scleral indentation

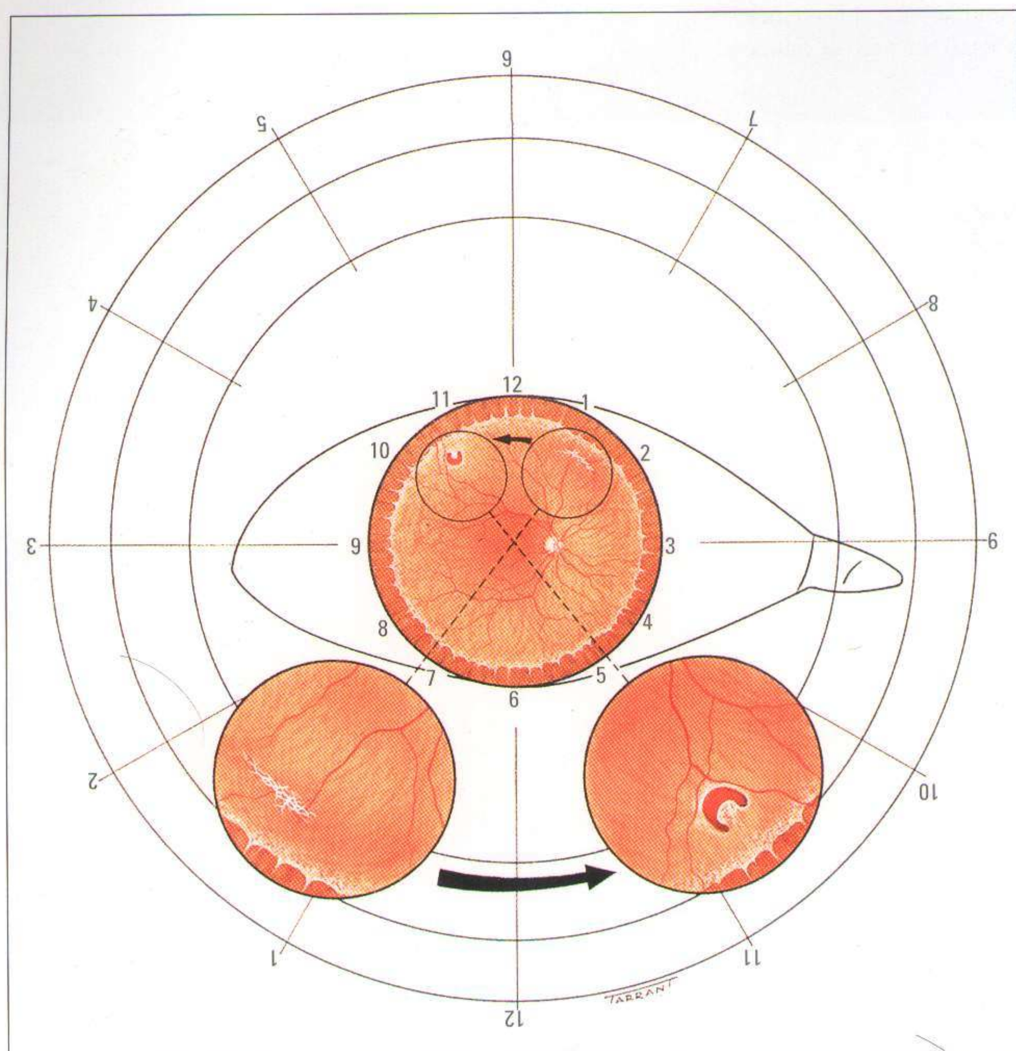


Fig. 12.14
Technique of drawing retinal lesions (*see text*)

of the upper eyelid at the margin of the tarsal plate (Fig. 12.12a).

2. With the indentor in place, the patient is asked to look up; the indentor is simultaneously advanced into the anterior orbit parallel with the globe (Fig. 12.12b).
3. The examiner's eyes should be aligned with the condensing lens and the indentor, with which gentle pressure is exerted. The indentation is observed as a mound in the fundus (Fig. 12.13). The indentor should be maintained tangential to the globe because perpendicular indentation is uncomfortable.
4. The indentor is moved to adjacent parts of the fundus, ensuring that the examiner's eyes, condensing lens and indentor are all in a straight line.

Fundus drawing

1. Technique. Because the image seen with the indirect ophthalmoscope is vertically inverted and laterally reversed, the top of the chart is placed towards the patient's feet (upside down). In this way the inverted position of the chart in relation to the patient's eye corresponds to the inverted image of the fundus. For example (Fig. 12.14), a U-tear at

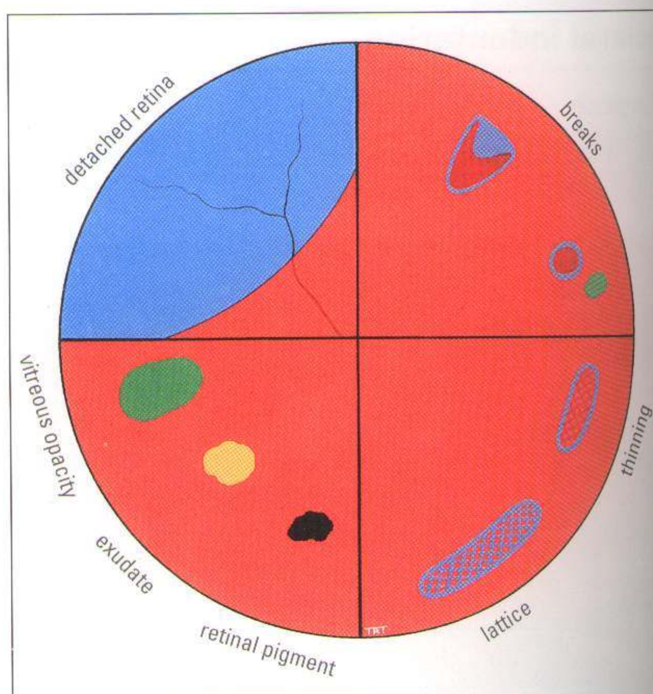


Fig. 12.15
Colour code for retinal drawing

11 o'clock in the eye will correspond to the 11 o'clock position on the chart. The same applies to the area of lattice degeneration between 1 and 2 o'clock.

2. **Colour code** (Fig. 12.15)
   - The boundaries of the RD are delineated, starting at the optic disc and then extending to the periphery.
   - Detached retina is drawn in blue and flat retina in red.
   - Retinal veins are drawn in blue but arteries are not drawn.
   - Retinal breaks are drawn in red with blue outlines; the flap of a retinal tear is drawn in blue.
   - Thin retina is indicated by red hatchings outlined in blue; lattice degeneration is shown as blue hatchings outlined in blue; retinal pigment as black; retinal exudates yellow; and vitreous opacities (including blood) green.

Goldmann triple-mirror examination

Goldmann triple-mirror

This consists of four parts (Fig. 12.16):

1. **The central part** provides a 30° view of the posterior pole.
2. **The equatorial mirror** (largest and oblong shaped) enables visualization from 30° to the equator.
3. **The peripheral mirror** (intermediate in size and square shaped) enables visualization between the equator and the ora serrata.
4. **The gonioscopic mirror** (smallest and dome shaped) may be used for visualizing the extreme retinal periphery and pars plana. It is therefore apparent that the smaller the mirror the more peripheral the view.

NB: The central part affords an upright virtual image of the posterior pole. With regard to the three mirrors:

- The mirror should be positioned opposite the area of the fundus to be examined.

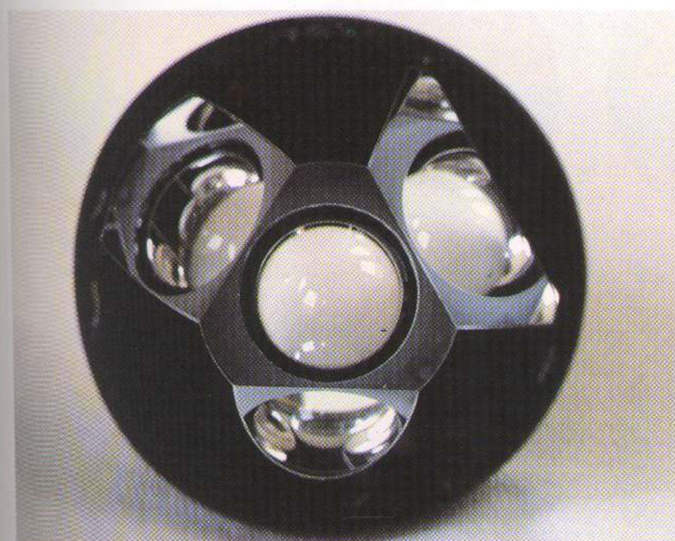


Fig. 12.16
Goldmann triple-mirror

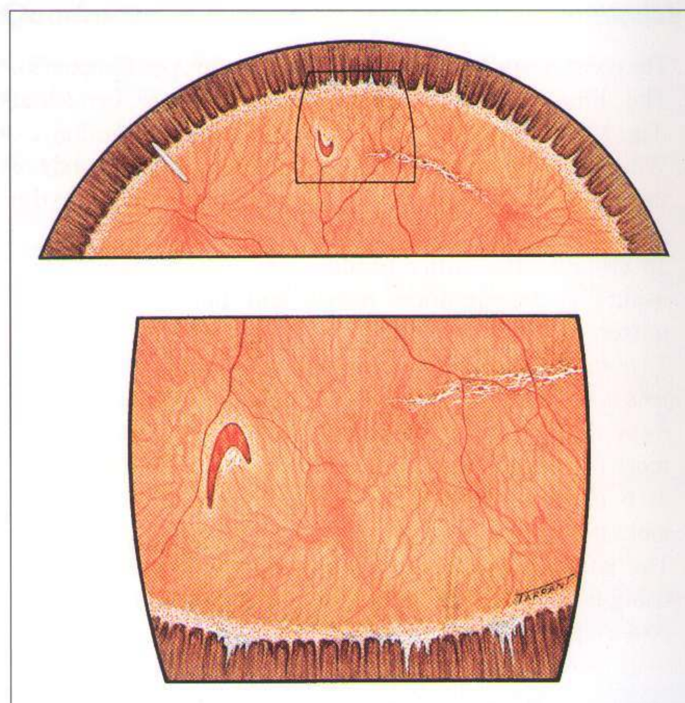


Fig. 12.17
(Top) U-tear left of 12 o'clock and an island of lattice degeneration right of 12 o'clock; **(bottom)** same lesions seen with the triple mirror positioned at 6 o'clock

- When viewing the vertical meridian, the image is upside down (Fig. 12.17).
- When viewing the horizontal meridian, the image is laterally reversed.



Fig. 12.18
Illumination column tilted and positioned right of centre to view the oblique meridians at 1.30 o'clock and 7.30 o'clock

Technique

1. The contact lens is inserted as for gonioscopy (*see* Chapter 9).
2. The illumination column should always be tilted (Fig. 12.18) except when viewing the vertical meridia.
3. When viewing sectors of the peripheral retina the axis of the beam is rotated so that it is always at right angles to the mirror.
4. To visualize the entire fundus the lens is rotated 360° using first the equatorial mirror and then the peripheral mirror.
5. To obtain a more peripheral view of any given sector the lens is tilted to the opposite side and the patient asked to move the eyes to the same side. For example, to obtain a more peripheral view of the 12 o'clock meridian (mirror at 6 o'clock) the lens is tilted down and the patient looks up.
6. The vitreous cavity is viewed through the central lens using both a horizontal and a vertical slit beam, and the posterior pole is then examined.

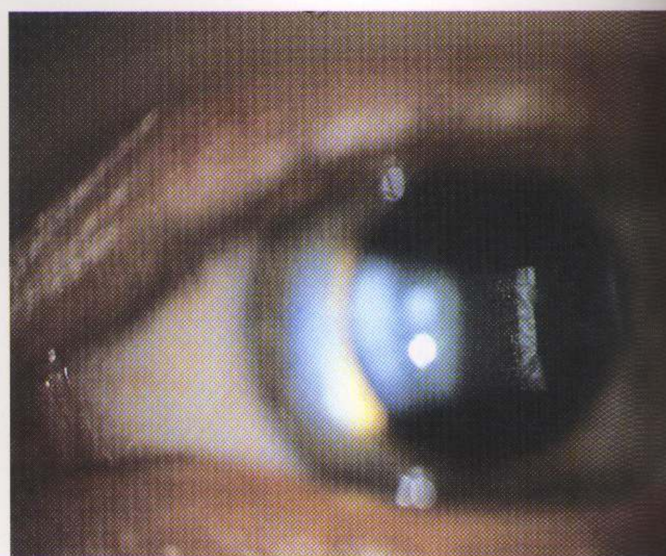


Fig. 12.19
Posterior vitreous detachment

Indirect slit-lamp biomicroscopy

This method utilizes high-power lenses (commonly +90 D and +78 D) designed to obtain a wide field of view. The lenses are used in a similar manner to an ordinary indirect ophthalmoscope lens and the image is likewise vertically inverted and laterally reversed.

Technique

1. The slit beam is adjusted to about one-quarter its full round diameter.
2. The illumination angle is set coaxially with the slit-lamp viewing system.
3. The lens is interposed in the slit beam immediately in front of the patient's eye (*see* Fig. 13.6).
4. The red reflex is identified, following which the microscope is pulled back, until the fundus comes into view.
5. The fundus is scanned, executing horizontal and vertical adjustment of the slit-lamp, while holding the lens still.
6. The width of the beam may be increased to obtain a larger field of view.
7. The magnification may be increased for greater detail as necessary.
8. To visualize peripheral retina the patient is asked to assume appropriate positions of gaze as with standard indirect ophthalmoscopy.



Fig. 12.20
Weiss ring (Courtesy of V. Tanner)

Interpretation of findings

- The normal vitreous in a young individual appears homogeneous with the same density throughout.
- The central vitreous cavity may contain optically empty spaces (lacunae). The condensed lining of a large cavity may be mistaken for a detached posterior hyaloid surface (pseudo-PVD).
- In eyes with PVD the detached posterior hyaloid surface can be seen (Fig. 12.19).
- A Weiss ring (Fig. 12.20) is an annular opacity representing a ring of glial tissue detached from the margin of the optic disc; it is virtually pathognomonic of PVD.
- Pigment cells ('tobacco dust') in the anterior vitreous in a patient complaining of sudden onset of flashing lights and floaters are strongly suggestive of a retinal tear (Fig. 12.21). A careful examination of the peripheral retina (particularly superiorly) is mandatory. The cells represent macrophages containing shed RPE cells.
- Numerous small opacities within the anteriorly displaced gel or in the retrohyaloid space are strongly suggestive of blood.

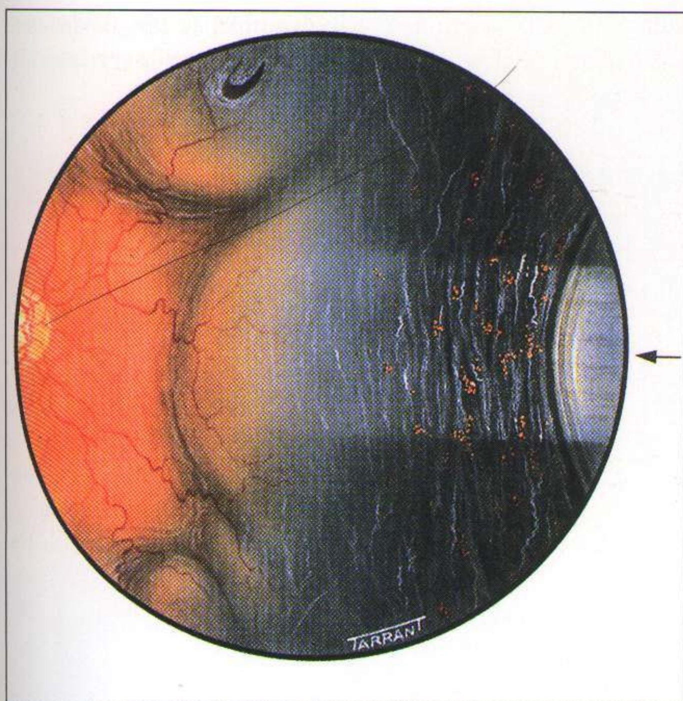


Fig. 12.21
Retinal detachment with a superior U-tear and tobacco dust

- Because of the wide field of view it may also be possible to visualize equatorial retinal tears.

Finding the primary retinal break

The primary break is the one deemed responsible for the RD, although further (secondary) breaks may also be present. Identifying the primary break is of paramount importance and aided by the following considerations.

Quadrantic distribution

- About 60% are in the upper temporal quadrant.
- About 15% are in the upper nasal quadrant.
- About 15% are in the lower temporal quadrant.
- About 10% are in the lower nasal quadrant.

The upper temporal quadrant is therefore by far the most common site for retinal break formation and should be examined in great detail if a break cannot be detected initially.

> ***NB:*** About 50% of eyes with RD have more than one break; and in most cases these are located within 90° of each other.

Configuration of retinal detachment

SRF usually spreads in a gravitational fashion. The configuration of an RD is governed by anatomical limits (ora serrata and optic disc) and the location of the primary retinal break. If the primary break is located superiorly, the SRF first gravitates inferiorly on the same side as the break and then ascends on the opposite side. The likely location of the primary retinal break can therefore be predicted by analysing the configuration of the RD (Fig. 12.22).

- A shallow inferior RD in which the SRF is slightly higher on the temporal side points to a primary break on that side (Fig. 12.22a).
- A primary break located at 6 o'clock will cause an inferior RD with equal fluid levels (Fig. 12.22b).
- In a bullous inferior RD the primary break usually lies above the horizontal meridian (Fig. 12.22c).
- If the primary break is located in the upper nasal quadrant the SRF will revolve around the optic disc and then rise

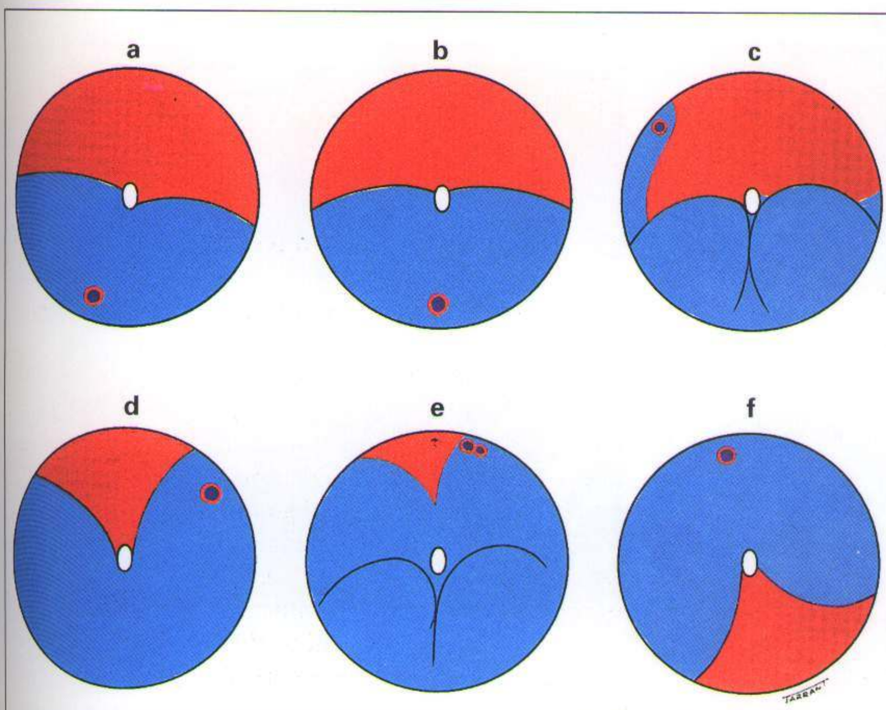


Fig. 12.22
Distribution of subretinal fluid in relation to the primary break

on the temporal side until level with the primary break (Fig. 12.22d).

- A subtotal RD with a superior wedge of attached retina points to a primary break located in the periphery nearest its highest border (Fig. 12.22e).
- When the SRF crosses the vertical midline above, the primary break is near 12 o'clock, the lower edge of the RD corresponding to the side of the break (Fig. 12.22f).

> ***NB:*** Diligent observation of the aforementioned principles prevents the treatment of secondary breaks while overlooking the primary break. Ensuring that the labelling of a break as 'primary' conforms to the configuration of the RD is therefore essential.

History

Although the quadrantic location of light flashes (photopsia) is of no value in predicting the location of the primary break, the quadrant in which the visual field defect first appeared may be of considerable value, since it corresponds to the area where the RD originated. For example, if the field defect started in the upper nasal quadrant the primary break is probably located in the lower temporal quadrant.

Ultrasonography

B-scan ultrasonography is useful in eyes with opaque media suspected of harbouring a retinal tear or RD. This applies particularly in the context of recent dense vitreous haemorrhage precluding visualization of the fundus. Under these circumstances ultrasonography may help to differentiate PVD (Fig. 12.23) from RD (Fig. 12.24). It may also be possible to detect the presence of a retinal tear in flat retina (Fig. 12.25). Dynamic ultrasonography, in which examination of intraocular structures is performed during lateral eye movements, is helpful in assessing the mobility of the



Fig. 12.23
Axial B-scan ultrasound showing intragel haemorrhage and posterior vitreous detachment (Courtesy of K. Nischal)



Fig. 12.24
Sagittal B-scan ultrasound showing inferior retinal detachment (Courtesy of K. Nischal)

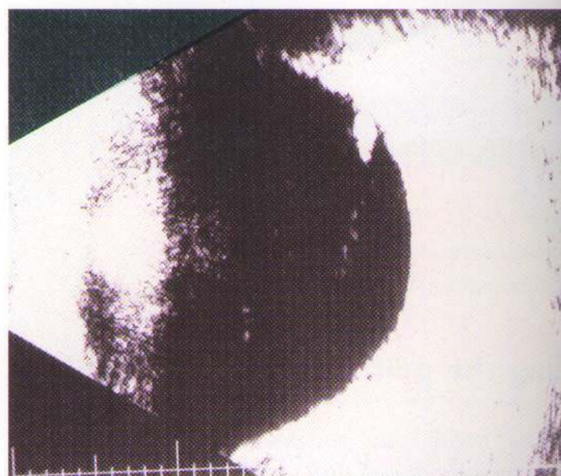


Fig. 12.25
Sagittal B-scan ultrasound showing a superior retinal tear associated with posterior vitreous detachment but a flat retina (Courtesy of K. Nischal)



Fig. 12.26
Sagittal B-scan ultrasound showing total retinal detachment associated with advanced proliferative vitreoretinopathy (triangular sign) (Courtesy of K. Nischal)

vitreous and retina in eyes with proliferative vitreoretinopathy (Fig. 12.26).

Pathogenesis of rhegmatogenous retinal detachment

Rhegmatogenous RD affects about 1:10,000 of the population each year and both eyes are eventually involved in about 10% of cases. The retinal breaks responsible for RD are caused by interplay between (a) *dynamic vitreoretinal traction* and (b) *predisposing degeneration* in the peripheral retina. Myopia may also play a significant role.

Dynamic vitreoretinal traction

Pathogenesis

Synchysis is a liquefaction of the vitreous gel (Fig. 12.27a). Some eyes with synchysis develop a hole in the thinned posterior vitreous cortex which overlies the fovea. Synchytic

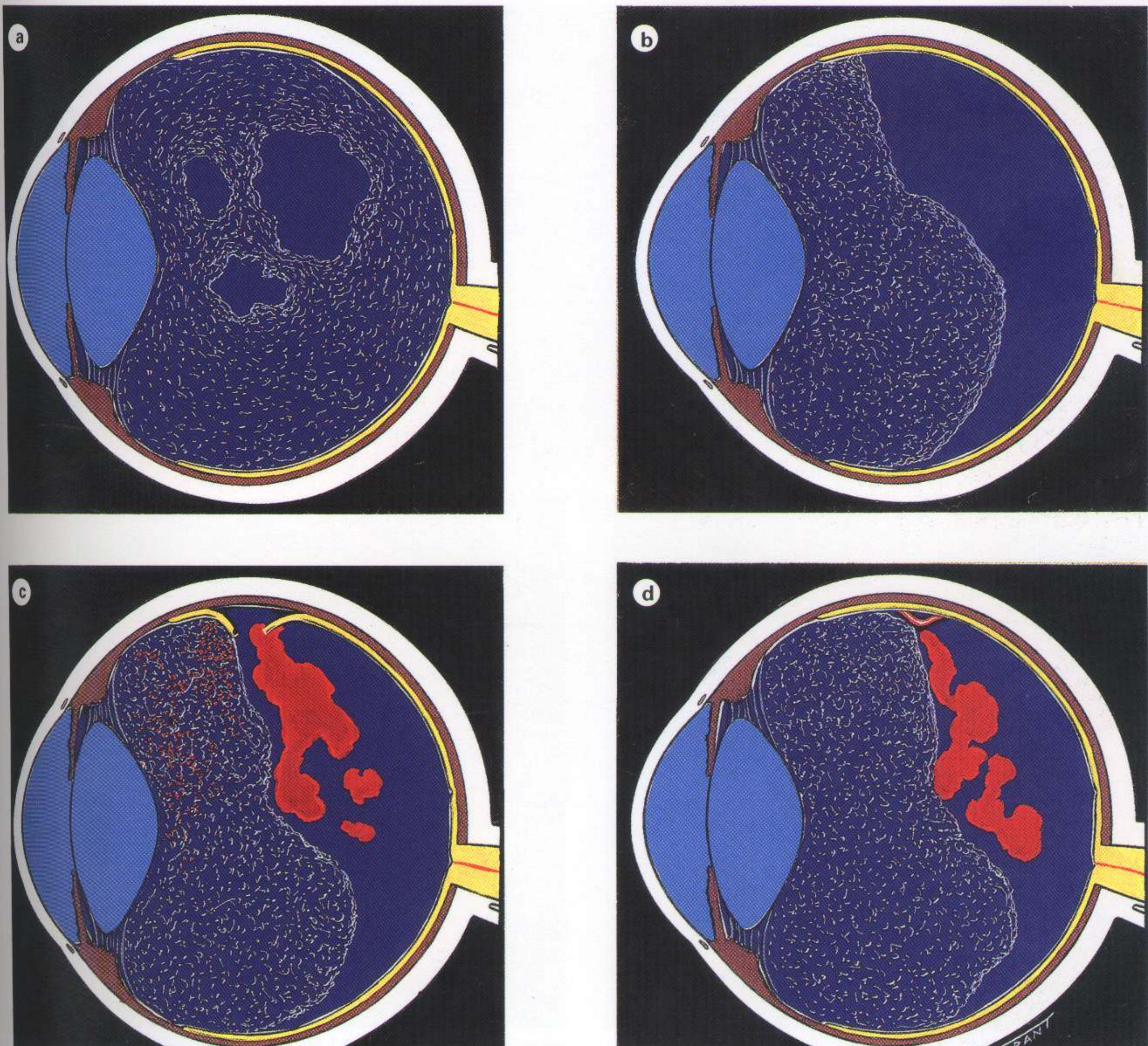


Fig. 12.27
(**a**) Synchysis; (**b**) uncomplicated posterior vitreous detachment; (**c**) retinal tear formation and vitreous haemorrhage; (**d**) avulsion of retinal blood vessel and vitreous haemorrhage

fluid from within the centre of the vitreous cavity passes through this defect into the newly formed retrohyaloid space. This process hydro-dissects the posterior vitreous surface from the internal limiting membrane (ILM) of the sensory retina as far forward as the posterior border of the vitreous base. The remaining solid vitreous gel collapses inferiorly and the retrohyaloid space is occupied entirely by synchytic fluid. This process is called acute rhegmatogenous PVD with collapse and will be referred to as acute PVD henceforth. The incidence of acute PVD increases with age and myopia.

Complications of acute PVD

These are dependent on the strength and extent of pre-existing vitreoretinal adhesions.

1. **No complications** (Fig. 12.27b) occur in most eyes, since vitreoretinal attachments are weak.
2. **Retinal tears** develop in about 10% of eyes as a result of traction at sites of abnormally strong vitreoretinal adhesions. Tears associated with acute PVD are usually symptomatic, U-shaped, located in the upper fundus (Fig. 12.28) and frequently associated with vitreous haemorrhage resulting from rupture of peripheral retinal blood vessels (*see* Fig. 12.27c). After a tear has formed, the synchytic retrohyaloid fluid has direct access to the subretinal space and unless the tear is treated prophylactically by photocoagulation or cryotherapy the risk of RD is high.
3. **Avulsion** of a peripheral retinal blood vessel resulting in vitreous haemorrhage in the absence of retinal tear formation (*see* Fig. 12.27d).

Predisposing peripheral retinal degenerations

About 60% of all breaks develop in areas of the peripheral retina that manifest specific changes. These lesions may be associated with spontaneous breakdown of pathologically thin retinal tissue to cause a retinal hole, or may predispose to retinal tear formation in eyes with acute PVD. Retinal holes are usually smaller than tears and carry a lower risk of RD.

Lattice degeneration

This is present in about 8% of the general population and in about 40% of eyes with RD. It is an important cause of RD in young myopes. Lattice-like lesions are frequently found in patients with Marfan syndrome, Stickler syndrome and Ehlers–Danlos syndrome, all of which are associated with an increased risk of RD.

1. **Signs**
   a. ***Typical*** lattice consists of sharply demarcated, circumferentially orientated, spindle-shaped areas of retinal thinning (Fig. 12.29), most frequently located between the equator and the posterior border of the vitreous base. Lattice is characterized by discontinuity of the internal limiting membrane, with variable atrophy of the underlying sensory retina. The lesions are usually bilateral and more often located in the temporal than nasal half of the fundus, and superiorly rather than inferiorly. A characteristic feature is an arborizing network of tiny white lines within the islands which may be associated with retinal pigment epithelial

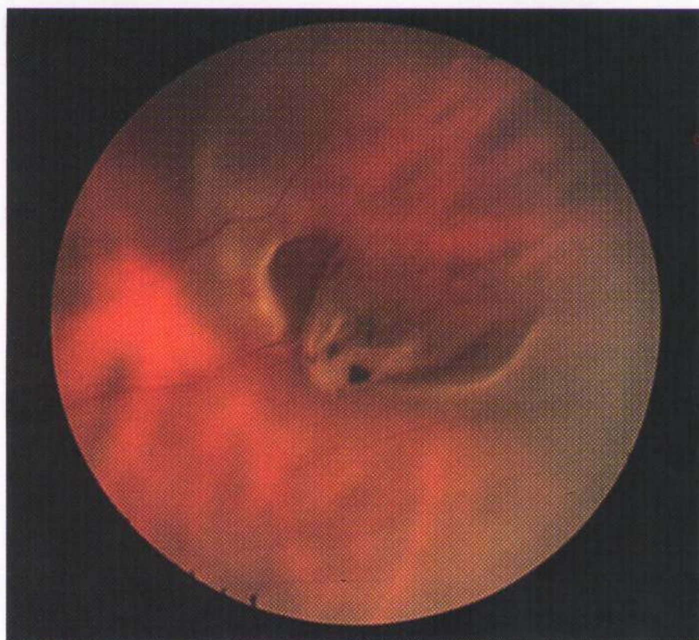


Fig. 12.28
Superior U-shaped tear associated with acute posterior vitreous detachment (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*, Criterion Press, Torrence California, 1982)

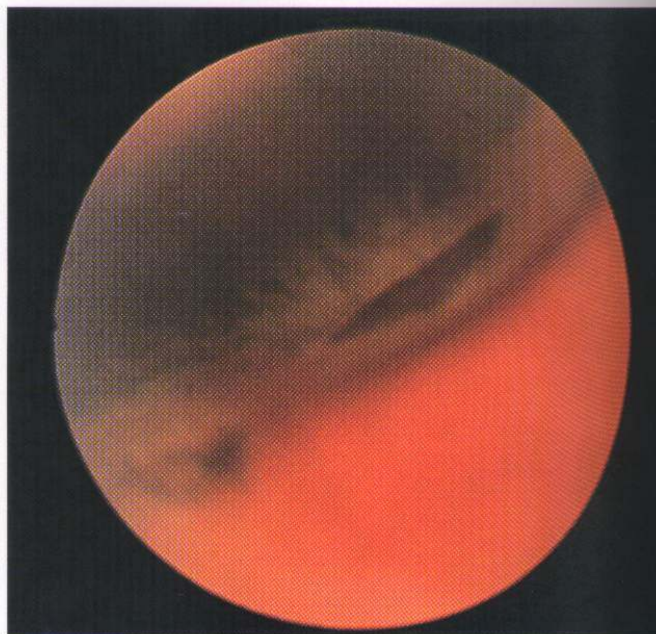


Fig. 12.29
Lattice degeneration forming a shallow crater (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence, California, 1982)



Fig. 12.30
Lattice degeneration with white lines and retinal pigment epithelial changes

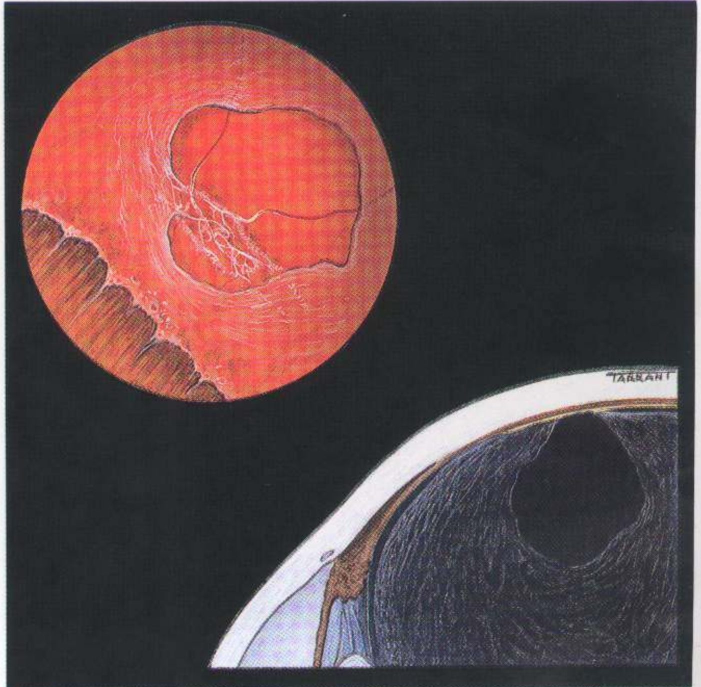


Fig. 12.32
Vitreous changes associated with lattice degeneration

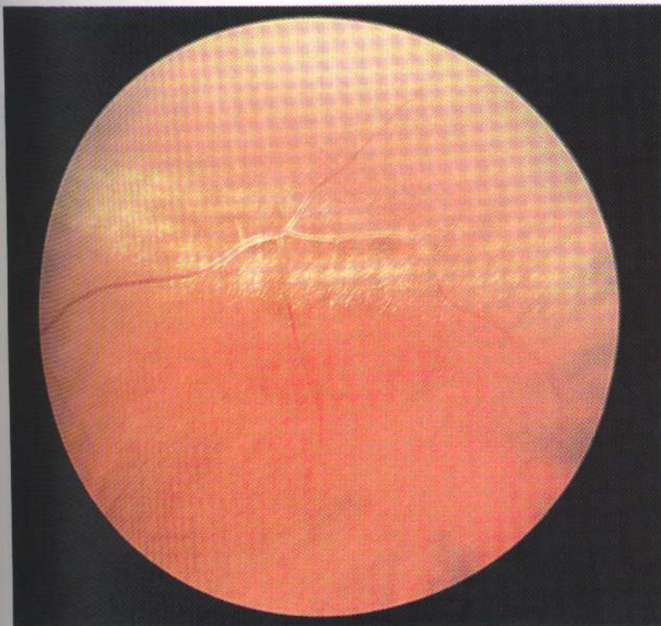


Fig. 12.31
Lattice degeneration with snowflakes (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence California, 1982)

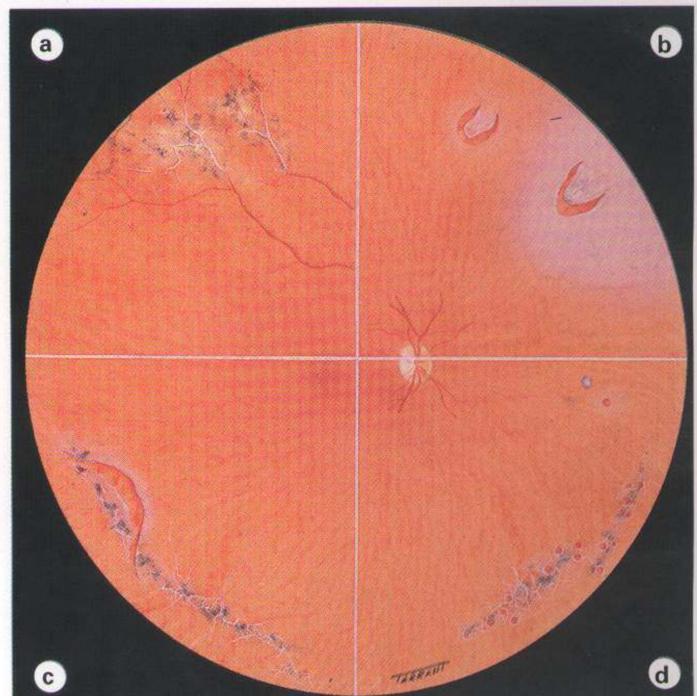


Fig. 12.33
(**a**) Atypical radial lattice degeneration; (**b**) lattice degeneration on the flap of a U-tear; (**c**) tractional tear along the posterior margin of lattice degeneration; (**d**) small round holes in lattice degeneration

changes (Fig. 12.30). Some lattice lesions may be associated with 'snowflakes' (remnants of degenerate Müller cells) (Fig. 12.31). The vitreous overlying an area of lattice is synchytic but the vitreous attachments around the margin of the lesion are exaggerated (Fig. 12.32).

b. ***Atypical*** lattice is characterized by radially orientated lesions continuous with peripheral blood vessels which may extend posterior to the equator (Fig. 12.33a). This type typically occurs in patients with Stickler syndrome.

2. Complications

- No complications are encountered in most patients, even in the presence of small holes which are frequently found within islands of lattice (Fig. 12.34, *see* Fig. 12.33d).

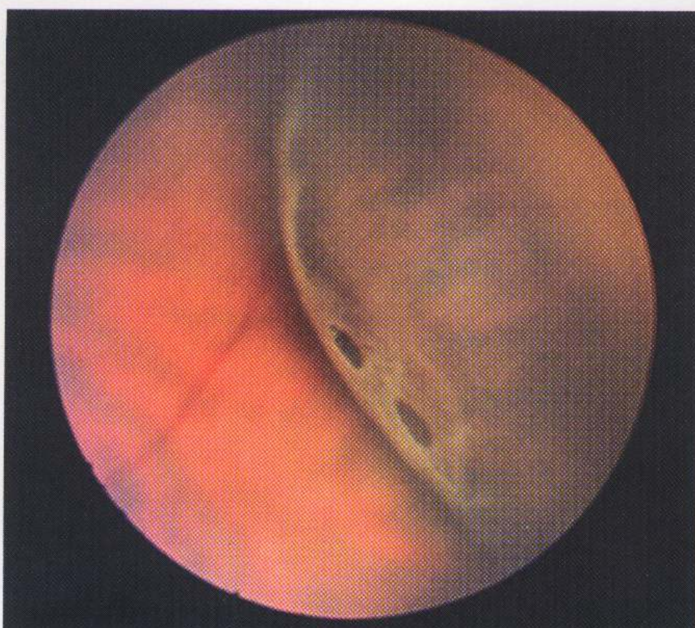


Fig. 12.34
Holes within lattice degeneration (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence California, 1982)



Fig. 12.36
Snailtrack degeneration seen on scleral indentation (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence California, 1982)

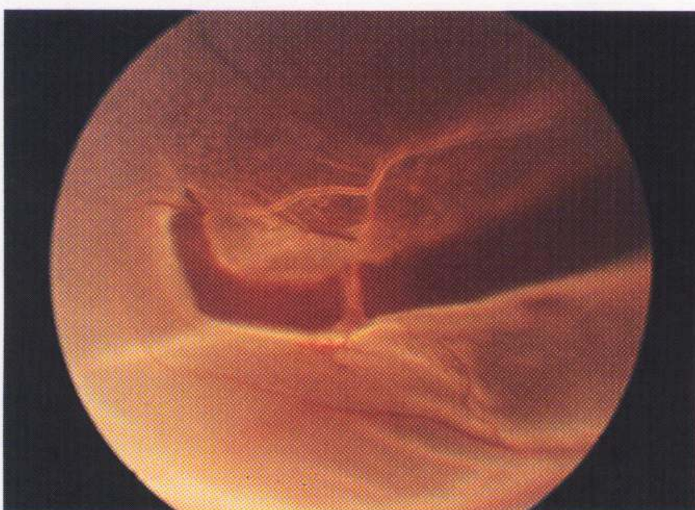


Fig. 12.35
Large tear associated with lattice degeneration (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence California, 1982)

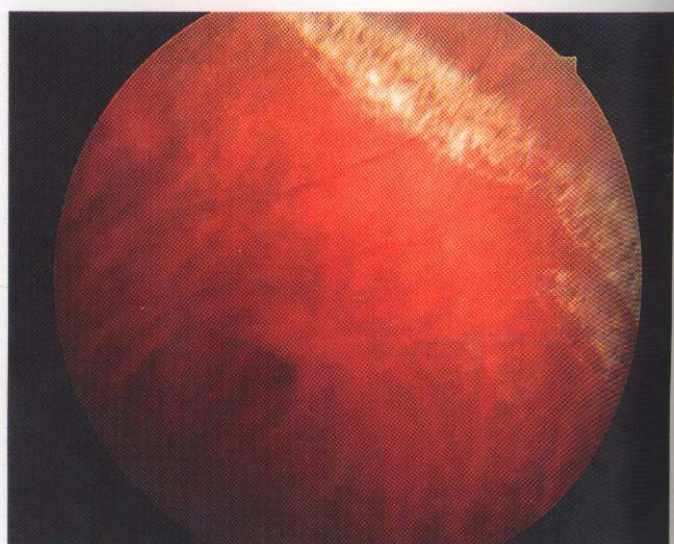


Fig. 12.37
Holes within snailtrack degeneration

> ***NB:*** RD associated with atrophic holes may occasionally occur, particularly in young myopes. In these patients it may not be preceded by symptoms of acute PVD (photopsia and floaters) and the SRF usually spreads slowly.

- RD due to tractional tears may occur in eyes with acute PVD (Fig. 12.35). Tractional tears typically develop along the posterior edge of an island of lattice (*see* Fig. 12.33c) as a result of dynamic traction at the site of tenacious vitreoretinal attachment. Occasionally a small island of lattice may be seen on the flap of a retinal tear (*see* Fig. 12.33b).

Snailtrack degeneration

1. **Signs**. Sharply demarcated circumferentially orientated bands of tightly packed 'snowflakes', which give the peripheral retina a white frost-like appearance. They are usually longer than islands of lattice (Fig. 12.36). Although snailtrack degeneration is associated with overlying vitreous liquefaction, marked vitreous traction at the posterior border of the lesions is seldom present, so that tractional U-tears rarely occur.
2. **Complications** include hole formation (Fig. 12.37) which may result in RD.

Degenerative retinoschisis

Retinoschisis is a splitting of the sensory retina into two layers: an outer (choroidal layer) and an inner (vitreous layer). The two main types are (a) *degenerative* and (b) *congenital* (*see* Chapter 15). Degenerative retinoschisis is present in about 5% of the population over the age of 20 years and is particularly prevalent in hypermetropes (70% of patients are hypermetropic) and is nearly always asymptomatic.

1. Classification

a. ***Typical***, in which the split occurs at the outer plexiform layer.

b. ***Reticular***, which is less common, in which the split is at the level of the nerve fibre layer.

2. Signs

- Early changes usually involve the extreme inferotemporal periphery of both fundi, appearing as an exaggeration of microcystoid degeneration with a smooth elevation of the retina (Fig. 12.38).
- Progression may occur circumferentially until the entire fundus periphery is involved. The typical type usually remains anterior to the equator although the reticular type may spread beyond the equator (Fig. 12.39).
- The surface of the inner layer may show 'snowflakes' as well as sheathing or 'silver-wiring' of blood vessels and the schisis cavity may be bridged by torn grey-white tissue (Fig. 12.40).
- The outer layer has a beaten-metal appearance and shows the phenomenon of 'white-with-pressure'.

NB: Unlike RD, retinoschisis is immobile.

3. Complications

- No complications occur in most cases and the condition is innocuous.

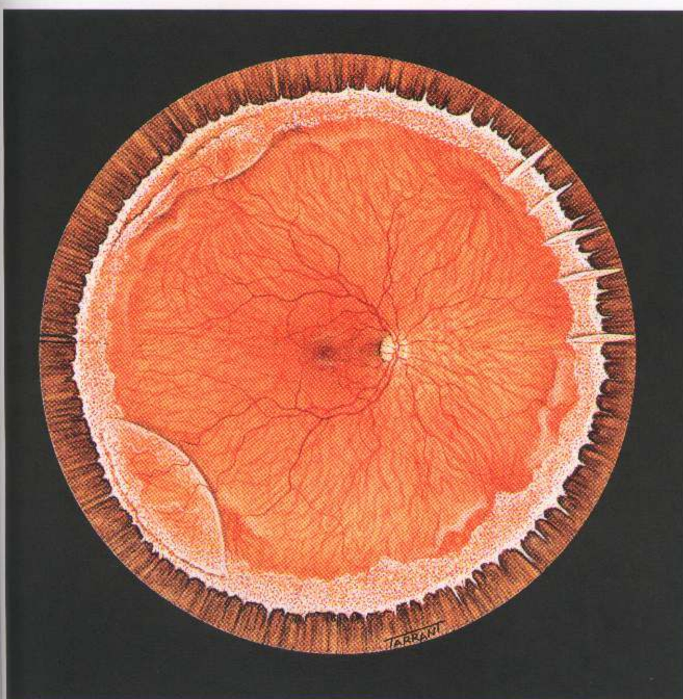


Fig. 12.38
Circumferential microcystoid degeneration and degenerative retinoschisis in the inferotemporal and superotemporal quadrants

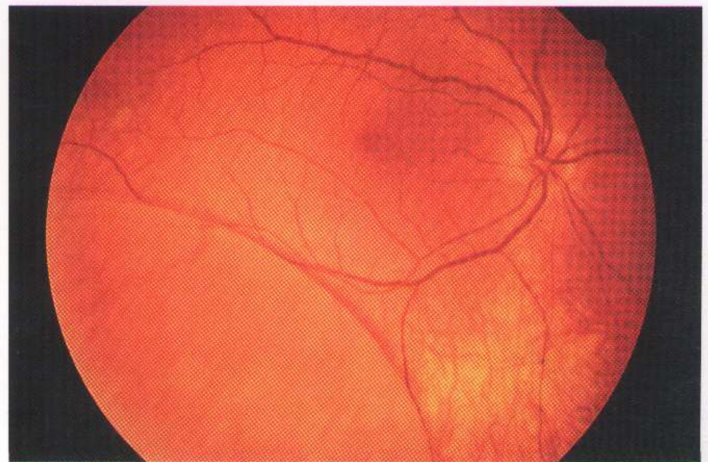


Fig. 12.39
Degenerative retinoschisis extending posterior to the equator

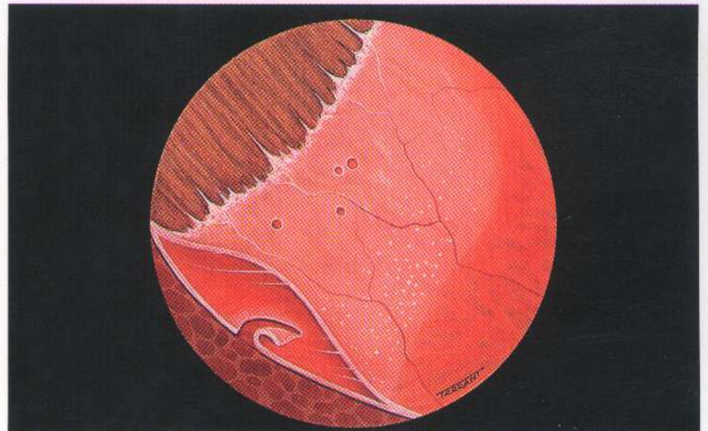


Fig. 12.40
Degenerative retinoschisis with holes in both layers; snowflakes and 'silver-wiring' of blood vessels on the inner layer; the cavity is bridged by torn grey-white tissue

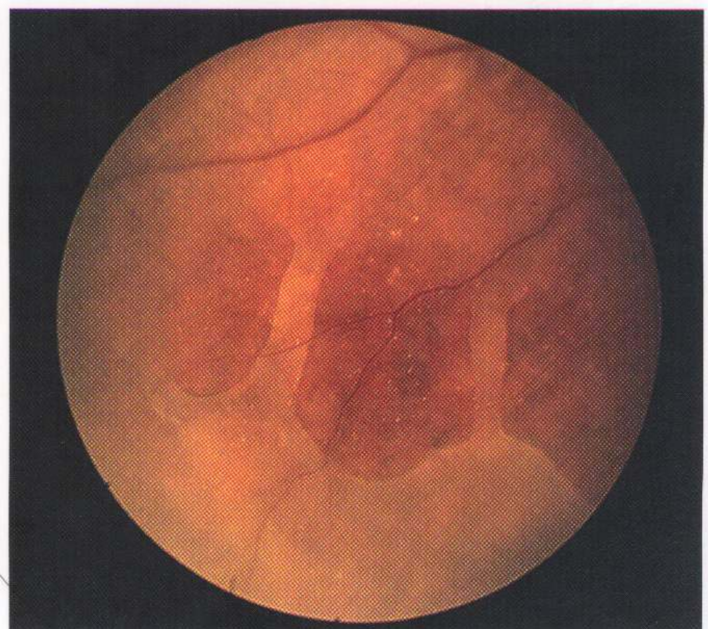


Fig. 12.41
Degenerative retinoschisis with snowflakes and outer layer breaks (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence California, 1982)

- Breaks may develop in the reticular type. Inner layer breaks are small and round (*see* Fig. 12.40), while the less common outer layer breaks are usually larger, with rolled edges and located behind the equator (Figs 12.41, 12.42a).
- RD is very rare, but may develop in eyes with breaks in both layers. Eyes with only outer layer breaks do not as a rule develop RD because the fluid within the schisis cavity is viscous and does not pass readily into the subretinal space. Rarely, however, the schisis fluid loses its viscosity and passes through the break into the subretinal space, giving rise to a localized detachment of the outer retinal layer which is usually confined to the area of retinoschisis (Fig. 12.42b).
- Vitreous haemorrhage is uncommon.

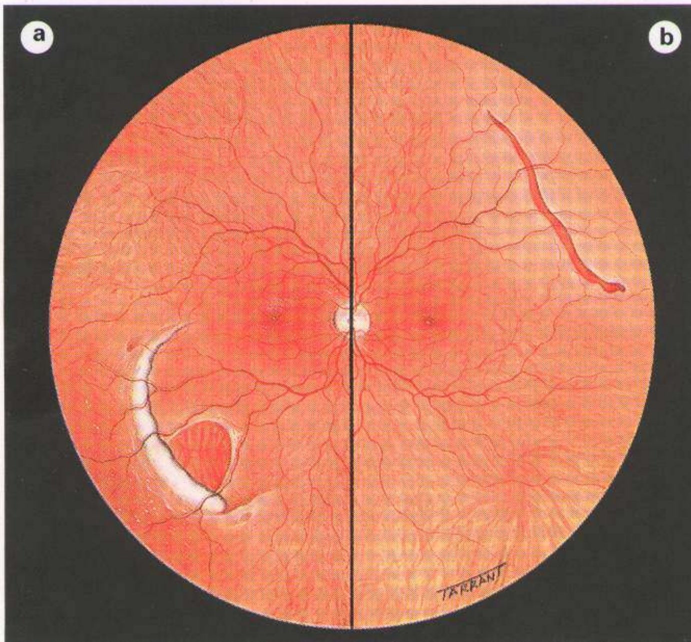


Fig. 12.42
Degenerative retinoschisis. (**a**) Large breaks in both layers; (b) linear break in the outer layer associated with a localized retinal detachment

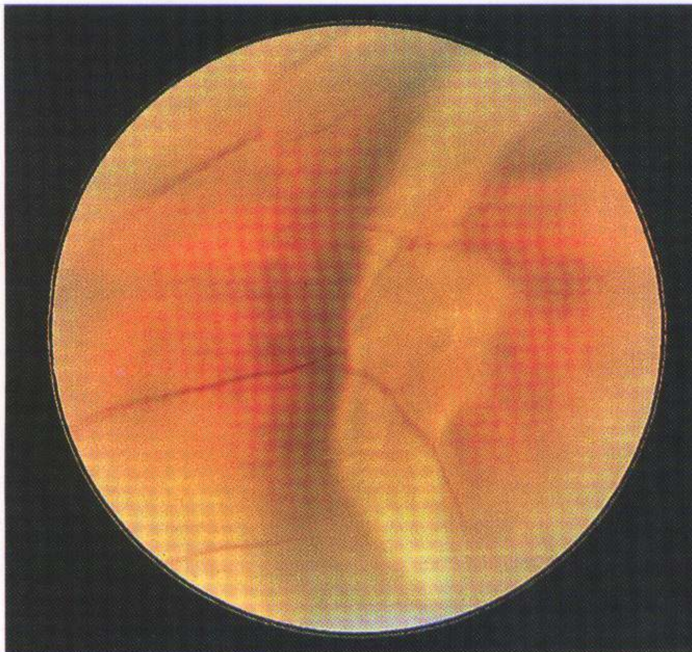


Fig. 12.43
White-with-pressure (Courtesy of N.E. Byer, *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence, California, 1982)

'White-without-pressure'

1. Signs

a. 'White-*with*-pressure' is a translucent grey appearance of the retina, induced by indenting the sclera (Fig. 12.43). Each area has a fixed configuration which does not change when the scleral indentor is moved to an adjacent area. It is often seen in normal eyes and may be observed along the posterior border of islands of lattice degeneration, snailtrack degeneration and outer layer of acquired retinoschisis.

b. 'White-*without*-pressure' has a similar appearance but is present without scleral indentation. On cursory examination a normal area of retina surrounded by

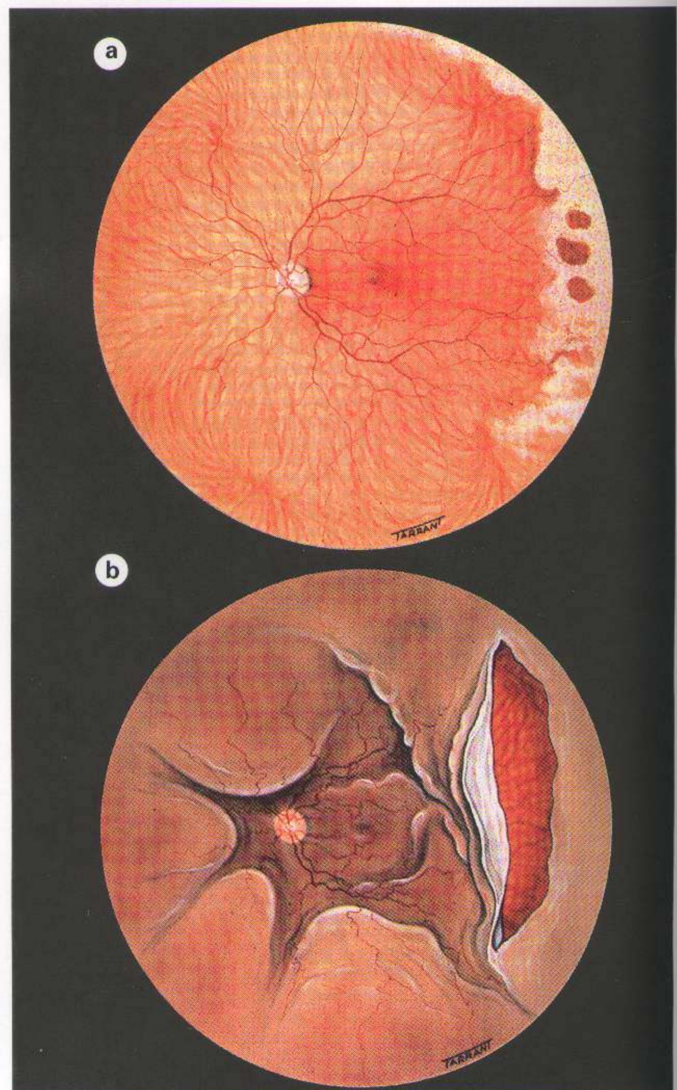


Fig. 12.44
(**a**) White-without-pressure with pseudo-holes; (**b**) total retinal detachment due to a giant tear

'white-*without*-pressure' may be mistaken for a flat retinal hole (Fig. 12.44a).

2. **Complications**. Giant tears occasionally develop along the posterior border of an area of 'white-*without*-pressure' (Fig. 12.44b).

Significance of myopia

Although myopes constitute 10% of the general population, over 40% of all RDs occur in myopic eyes. The higher the refractive error the greater the risk of RD. The following interrelated factors predispose the myopic eye to RD:

1. **Lattice degeneration** is more common in moderate myopes and may give rise to tears or holes.
2. **Snailtrack degeneration** is common in myopic eyes and may be associated with holes.
3. **Diffuse chorioretinal atrophy** may give rise to small holes in highly myopic eyes.
4. **Macular holes** may give rise to RD in highly myopic eyes.
5. **Vitreous degeneration** and PVD are more common.
6. **Vitreous loss** during cataract surgery, particularly if inappropriately managed, is associated with about a 15% incidence of subsequent RD in myopic eyes greater than 6 D; the risk is even higher if myopia is more than 10 D.
7. **Posterior capsulotomy** is associated with an increased risk of RD in myopic eyes.

Clinical features of retinal detachment

Rhegmatogenous retinal detachment

Symptoms

The classic premonitory symptoms reported in about 60% of patients with spontaneous rhegmatogenous RD are photopsia and vitreous floaters. After a variable period of time the patient notices a relative peripheral visual field defect which may progress to involve central vision.

1. **Photopsia** in eyes with acute PVD is probably caused by traction on the retina at sites of vitreoretinal adhesion. Cessation of photopsia is the result of either separation of the adhesion or complete tearing away of a piece of retina (operculum) at the site of adhesion. In eyes with PVD the photopsia may be induced by eye movements and is more noticeable in dim illumination. It tends to be projected into the temporal peripheral visual field and, unlike floaters, has no lateralizing value.
2. **Floaters** are moving vitreous opacities perceived when they cast a shadow upon the retina. Vitreous opacities in eyes with acute PVD are of the following three types:
   a. A solitary ring-shaped opacity representing the detached annular attachment to the margin of the optic disc (Weiss ring) (*see* Fig. 12.20).
   b. Cobwebs are caused by condensation of collagen fibres within the collapsed vitreous cortex.
   c. A sudden shower of minute red-coloured or dark spots usually indicates vitreous haemorrhage secondary to tearing of a peripheral retinal blood vessel.
3. **A visual field defect** secondary to RD is perceived as a dark curtain. In some patients it may not be present on waking in the morning due to spontaneous absorption of SRF, only to reappear later in the day. The quadrant of the visual field in which the defect first appears is useful in predicting the location of the primary retinal break (which will be in the opposite quadrant). Loss of central vision may be due either to involvement of the fovea by SRF or, less frequently, obstruction of the visual axis by a large upper bullous RD (Fig. 12.45).

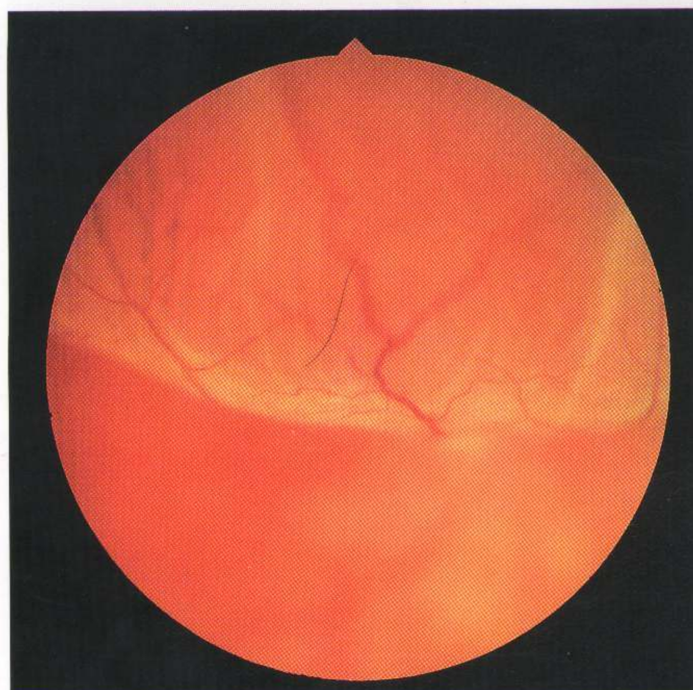


Fig. 12.45
Fresh superior bullous retinal detachment



Fig. 12.46
Tobacco dust and cobweb-like opacities associated with retinal detachment (Courtesy of V. Tanner)

General signs

- A Marcus Gunn pupil (relative afferent pupillary defect) is present in eyes with extensive RDs irrespective of the type.
- The intraocular pressure is usually lower than in the normal eye by about 5 mmHg.
- A mild anterior uveitis is very common.
- The anterior vitreous shows 'tobacco dust' (Fig. 12.46).
- Retinal breaks appear as red discontinuities in the retinal surface (Fig. 12.47).
- Retinal signs depend on the duration of RD and the presence or absence of proliferative vitreoretinopathy as described below.

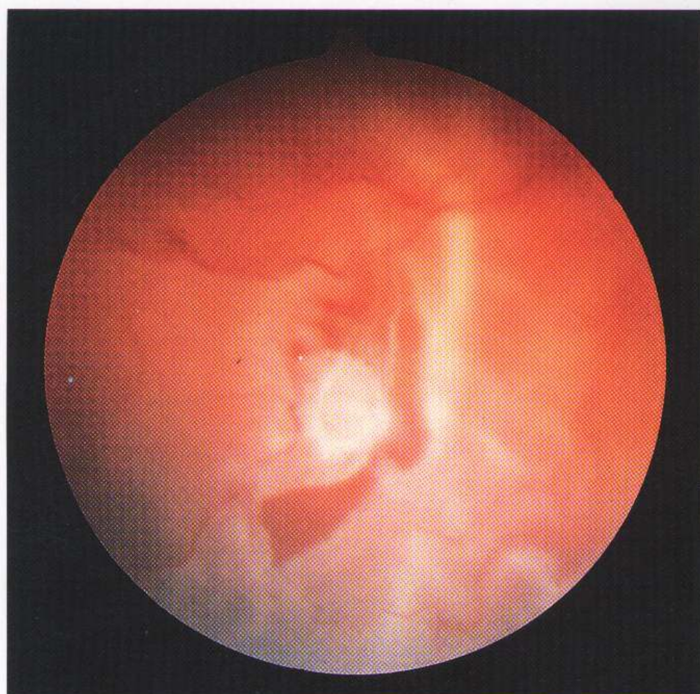


Fig. 12.47
Retinal tears in detached retina

Fresh retinal detachment

- The detached retina has a convex configuration and a slightly opaque and corrugated appearance due to intraretinal oedema. It undulates freely with eye movements (Fig. 12.48).
- There is loss of the underlying choroidal pattern and the retinal blood vessels appear darker than in flat retina, so that the colour contrast between venules and arterioles is less apparent.
- The SRF extends up to the ora serrata except in the rare cases caused by a macular hole, in which the SRF is initially confined to the posterior pole.
- A pseudo-hole is frequently seen if the posterior pole is detached.

> *NB:* This should not be mistaken for a true macular hole which may give rise to RD in highly myopic eyes or following blunt ocular trauma.

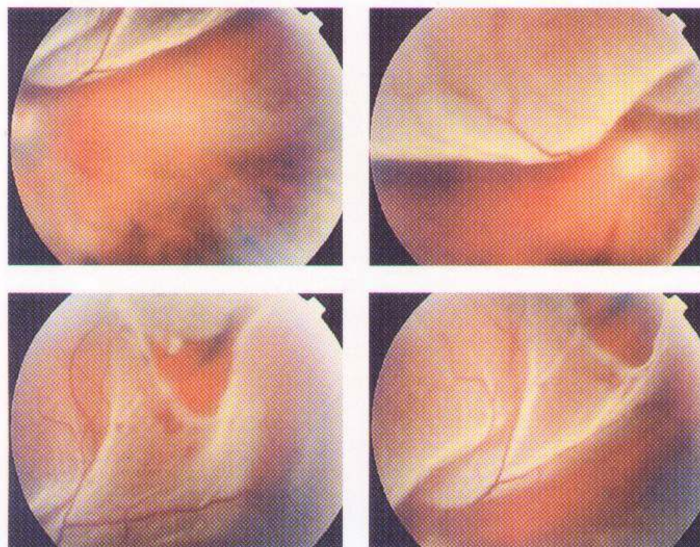


Fig. 12.48
Fresh retinal detachment due to a large U-shaped tear with mobile subretinal fluid (Courtesy of S. Milewski)

Old retinal detachment

The following are the main features of a long-standing rhegmatogenous RD which do not occur in any other type of RD irrespective of duration (Fig. 12.49).

- Retinal thinning secondary to atrophy, which must not be mistaken for retinoschisis.
- Secondary intraretinal cysts may develop if the RD has been present for over a year (Fig. 12.50).
- Subretinal demarcation lines (high-water marks) caused by proliferation of RPE cells at the junction of flat and

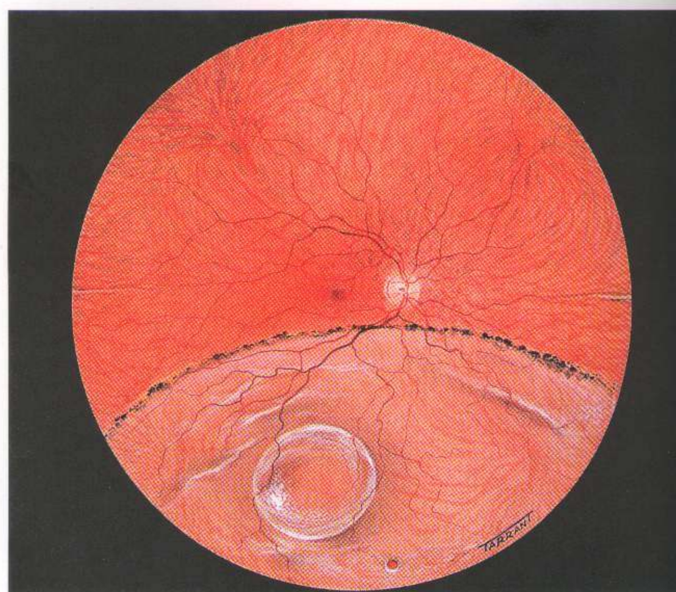


Fig. 12.49
Long-standing inferior retinal detachment associated with a secondary intraretinal cyst and a pigmented demarcation line

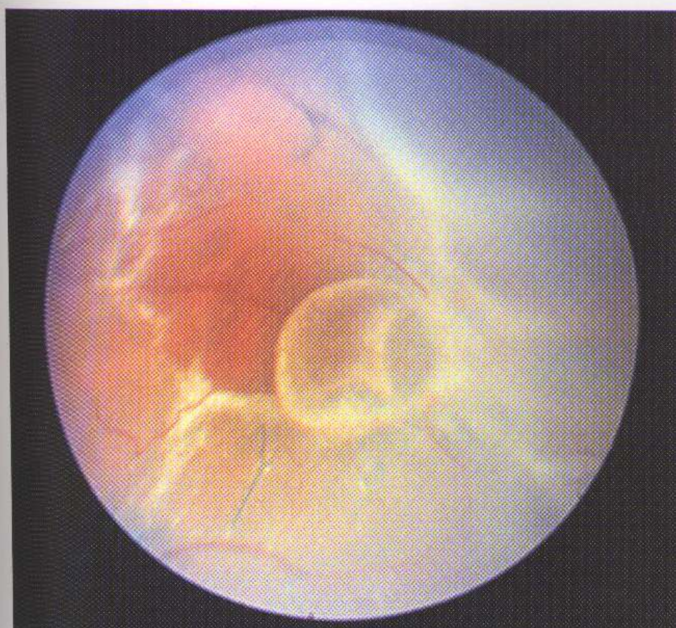


Fig. 12.50
Secondary intraretinal cyst (Courtesy of N.E. Byer, from *The Peripheral Retina in Profile, a Stereoscopic Atlas*. Criterion Press, Torrence, California, 1982)



Fig. 12.52
Retinal tears with rolled edges in grade B proliferative vitreoretinopathy

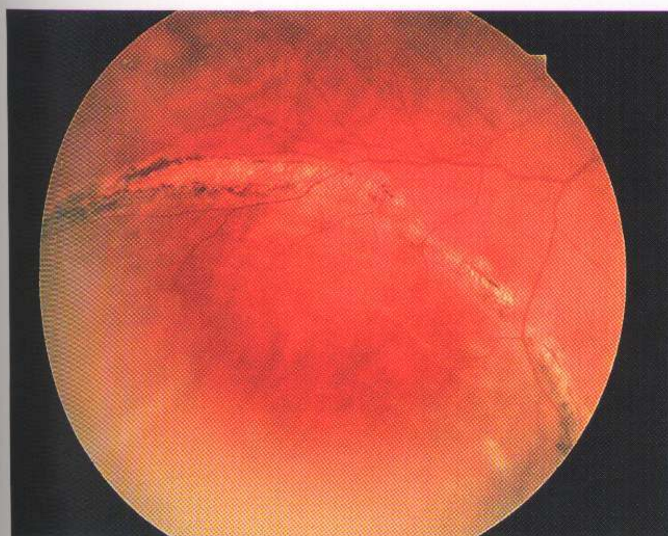


Fig. 12.51
Inferior retinal detachment demarcated by a pigmented line

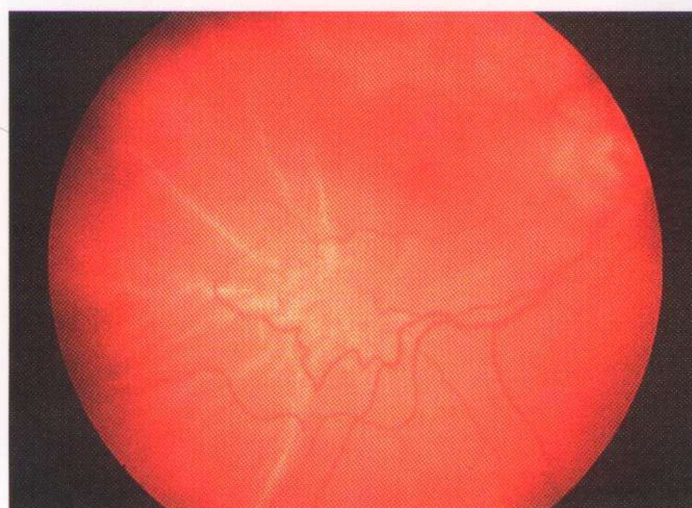


Fig. 12.53
Retinal wrinkling and vascular distortion in grade B proliferative vitreoretinopathy

detached retina are common and take about 3 months to develop (Fig. 12.51).

Proliferative vitreoretinopathy

Proliferative vitreoretinopathy (PVR) is caused by the proliferation and contraction of membranes on the inner retinal surface (epiretinal membranes), on the posterior surface of the detached hyaloid and occasionally also on the outer retinal surface (subretinal membranes). Severe postoperative contraction of these membranes is the most common cause of failure of RD surgery. The main clinical signs of PVR are retinal folds and rigidity so that retinal undulation induced by eye movements or scleral indentation is decreased according to severity. The classification of PVR is as follows:

1. **Grade A** (minimal) is characterized by diffuse vitreous haze, 'tobacco dust' and occasionally pigmented cells on the inferior retina.
2. **Grade B** (moderate) is characterized by retinal breaks with rolled irregular edges (Fig. 12.52), wrinkling of the inner retinal surface and tortuous blood vessels (Fig. 12.53), retinal stiffness and decreased mobility of vitreous gel. The epiretinal membranes responsible for these findings cannot be identified by indirect ophthalmoscopy, although they may be visualized on slit-lamp non-contact indirect ophthalmoscopy.

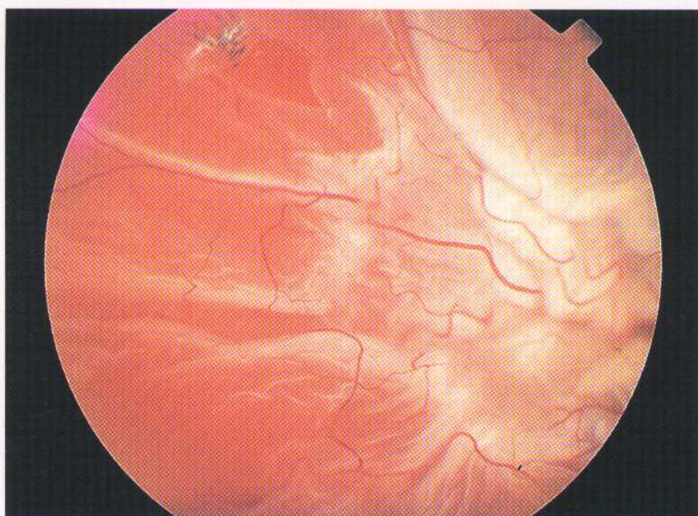


Fig. 12.54
Fixed retina folds in grade C proliferative vitreoretinopathy

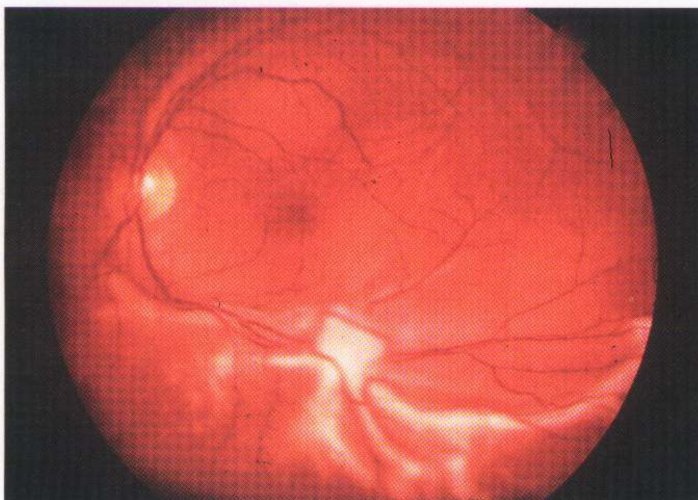


Fig. 12.55
Grade C type 1 (focal) proliferative vitreoretinopathy

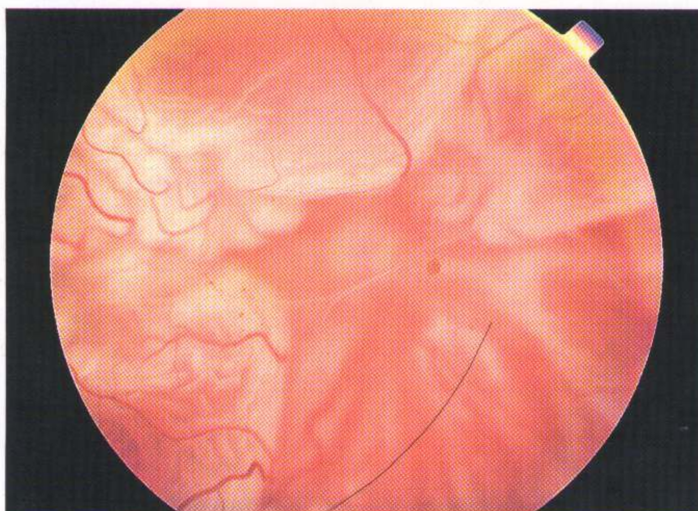


Fig. 12.56
Grade C type 2 (diffuse) proliferative vitreoretinopathy

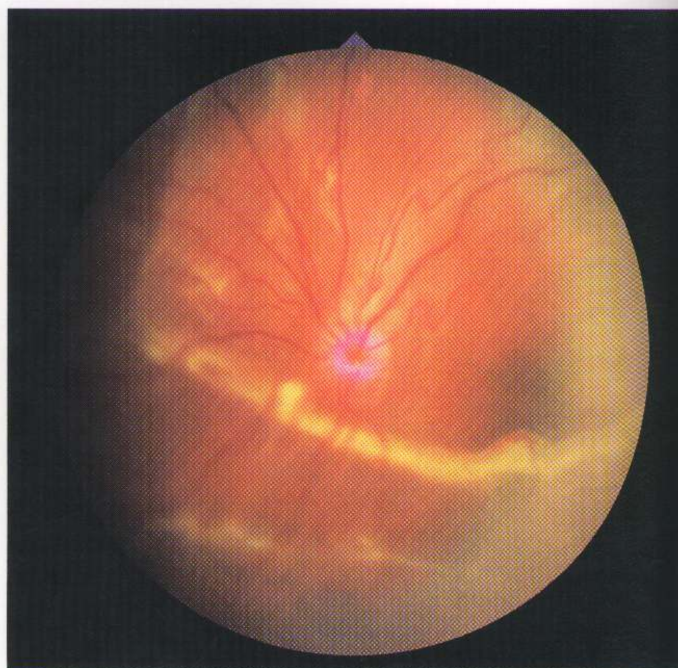


Fig. 12.57
Grade C type 3 (subretinal) proliferative vitreoretinopathy

3. **Grade C** (severe) is characterized by full-thickness rigid retinal folds (Fig. 12.54) with heavy vitreous condensation and strands. It may be anterior or posterior, the rough dividing line being the equator of the globe.
   a. ***The severity*** of proliferation is expressed by the number of clock hours of retina involved (1–12), although proliferations need not be contiguous.
   b. ***The type*** of contraction is further subdivided into: type 1 (focal) (Fig. 12.55), type 2 (diffuse) (Fig. 12.56), type 3 (subretinal) (Fig. 12.57), type 4 (circumferential) and type 5 (anterior displacement).

Tractional retinal detachment

1. **Symptoms**. Photopsia and floaters are usually absent because vitreoretinal traction develops insidiously and is not associated with acute PVD. The visual field defect usually progresses slowly and may become stationary for months or even years.
2. **Signs**
   - The detached retina is concave and breaks are absent (Fig. 12.58).
   - The SRF is shallower than in a rhegmatogenous RD and seldom extends to the ora serrata (Fig. 12.59).
   - The highest elevation of the retina occurs at sites of vitreoretinal traction.
   - Retinal mobility is severely reduced and shifting fluid is absent.

> ***NB:*** If a tractional RD develops a break it assumes the characteristics of a rhegmatogenous RD and progresses more quickly (combined tractional–rhegmatogenous RD) (Fig. 12.60).


Fig. 12.58
Inferior tractional retinal detachment in proliferative diabetic retinopathy


Fig. 12.60
Combined tractional–rhegmatogenous retinal detachment in proliferative diabetic retinopathy

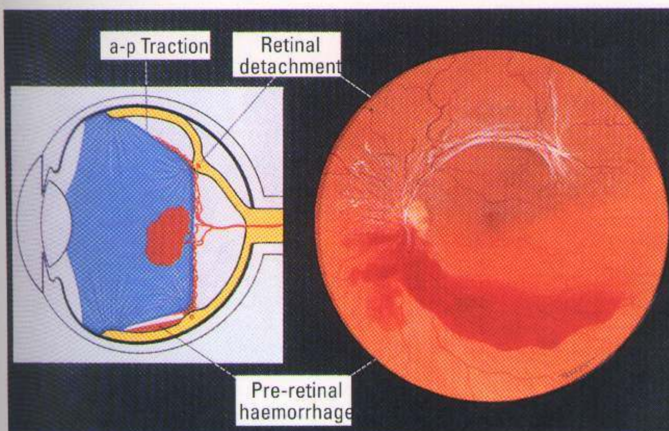
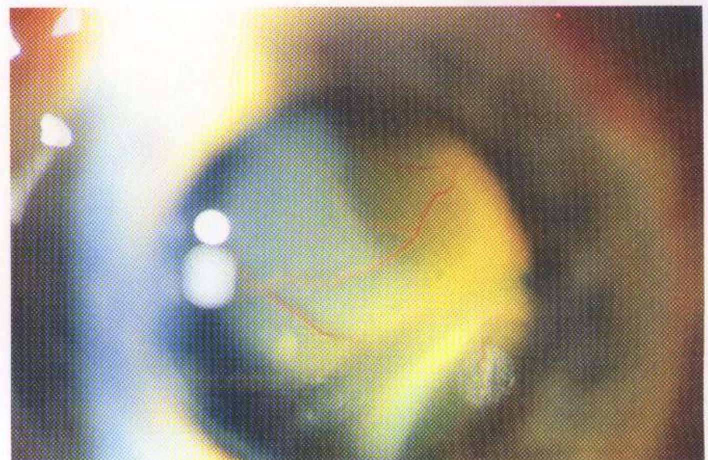


Fig. 12.59
Superior tractional retinal detachment in proliferative diabetic retinopathy


Fig. 12.61
Exudative retinal detachment

Exudative retinal detachment

1. **Symptoms**. Photopsiae are absent because there is no vitreoretinal traction although floaters may be present if there is associated vitritis. The visual field defect may develop suddenly and progress rapidly. In some cases of Harada disease both eyes may be involved simultaneously.
2. **Signs**
   - The detached retina is convex and retinal breaks are absent.
   - The surface is smooth rather than corrugated.
   - Occasionally the SRF is so deep that the RD may be seen on the slit-lamp without the aid of a lens (Fig. 12.61); the retina may even touch the back of the crystalline lens.
   - The detached retina is very mobile and exhibits the phenomenon of 'shifting fluid' in which SRF responds to the force of gravity and detaches the area of retina under which it accumulates. For example, in the upright position the SRF collects under the inferior retina but on assuming the supine position the inferior retina flattens and the SRF shifts posteriorly, detaching the macula and superior retina.
   - Scattered areas of subretinal clumping giving rise to characteristic 'leopard spots' may be seen after the detachment has resolved (Fig. 12.62).
   - The cause of the RD, such as a choroidal tumour (Fig. 12.63), may be apparent when the fundus is examined.

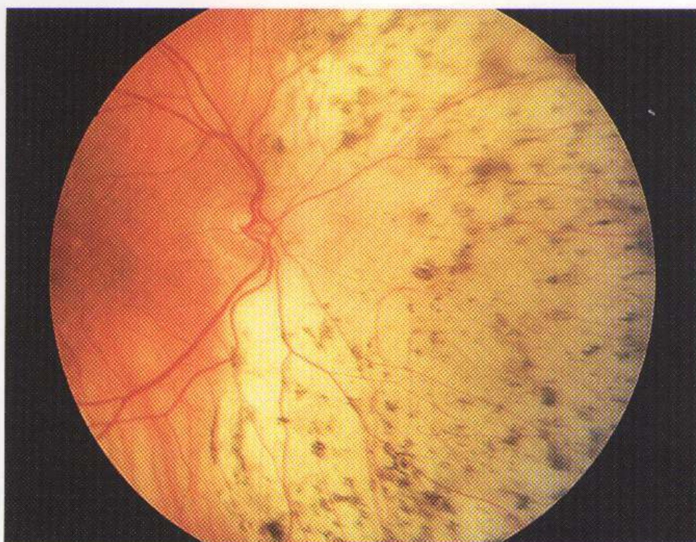


Fig. 12.62
'Leopard spot' pigmentation following resolution of exudative retinal detachment

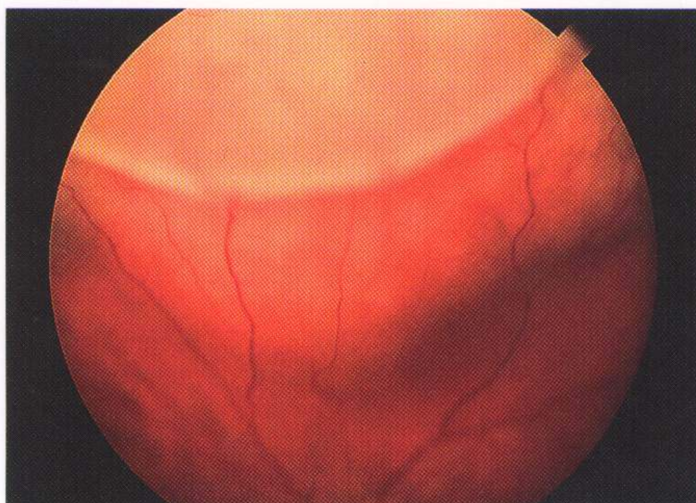


Fig. 12.63
Choroidal metastasis with overlying exudative retinal detachment (Courtesy of J. Shields and A. Singh)

Differential diagnosis of retinal detachment

Degenerative retinoschisis

1. **Symptoms**. Photopsia and floaters are absent because there is no vitreoretinal traction. A visual field defect is seldom observed because posterior spread is rare. If present it is absolute.
2. **Signs** (*see* Figs 12.38, 12.39 and 12.40)
   - The elevation is convex, smooth, thin and immobile.
   - The thin inner leaf of the schisis may be mistaken, on cursory examination, for an atrophic long-standing rhegmatogenous RD. However, demarcation lines and secondary cysts in the inner leaf are absent in retinoschisis.
   - Breaks may be present in one or both layers in eyes with reticular retinoschisis.

Choroidal detachment

1. **Symptoms**. Photopsia and floaters are absent because there is no vitreoretinal traction. A visual field defect may be present in an eye with a large choroidal detachment.
2. **Signs**
   - The intraocular pressure may be very low as a result of concomitant detachment of the ciliary body.
   - A choroidal detachment is a brown, convex, smooth, bullous elevation, which is relatively immobile (Fig. 12.64).
   - The peripheral retina and ora serrata may be seen without scleral indentation (Fig. 12.65).

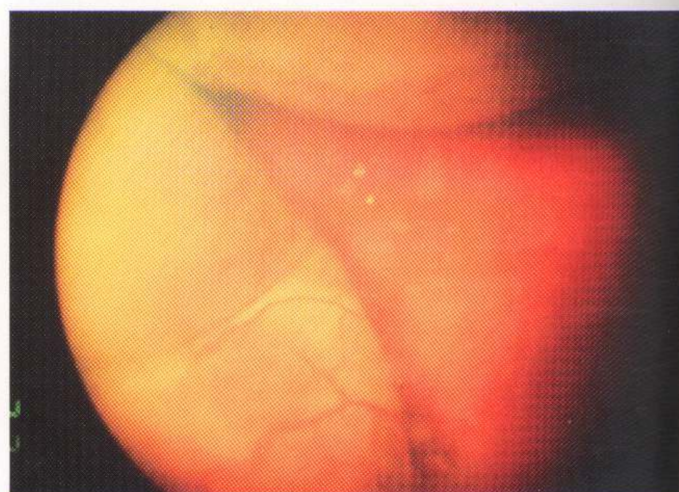


Fig. 12.64
Choroidal detachment

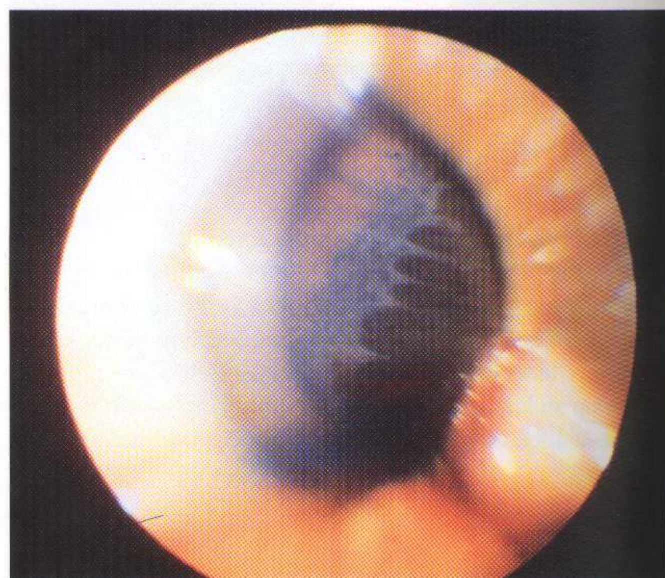


Fig. 12.65
Choroidal detachment with visibility of the pars plana (Courtesy of P. Morse)

- The elevations do not extend to the posterior pole because they are limited by the firm adhesion between the suprachoroidal lamellae and the sclera where the vortex veins enter their scleral canals.

Uveal effusion syndrome

The uveal effusion syndrome is a rare, idiopathic condition characterized by choroidal detachments associated with exudative RD (Fig. 12.66). Following resolution, the RPE frequently shows a characteristic residual mottling.

> *NB:* Uveal effusion may be mistaken for either RD complicated by choroidal detachment or ring melanoma of the anterior uvea.

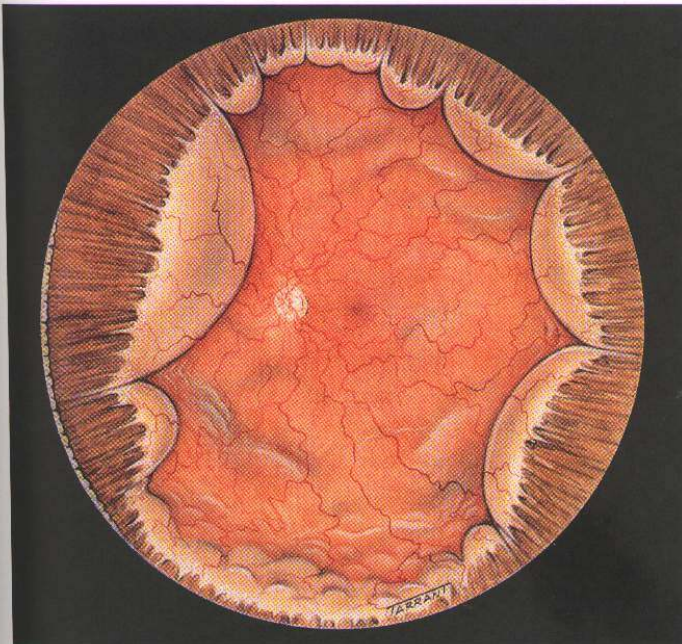


Fig. 12.66
Uveal effusion syndrome characterized by choroidal detachment and exudative retinal detachment

Prophylaxis of rhegmatogenous retinal detachment

Retinal breaks

Although, given the right circumstances, any retinal break may cause an RD, some are more dangerous than others. Important criteria in the selection of patients for prophylactic treatment can be divided into (a) *type of break* and (b) *other considerations*.

Type of break

1. **Tears** are more dangerous than holes because they are associated with dynamic vitreoretinal traction.
2. **Large breaks** are more dangerous than small ones because of increased access to the subretinal space.
3. **Symptomatic breaks** are more dangerous than those detected by chance because they are associated with dynamic vitreoretinal traction.
4. **Superior breaks** are more dangerous than inferior breaks because SRF is likely to spread more quickly.
5. **Equatorial breaks** are more dangerous than oral breaks, which seldom give rise to RD.
6. **A subclinical RD** constitutes a break surrounded by a very small amount of SRF. In some cases the SRF may spread and the RD become 'clinical' within a short period of time.
7. **Pigmentation** around a retinal break indicates that it is long-standing with a small risk of RD.

Other considerations

1. **Aphakic** patients are at increased risk of RD, particularly when surgery has been complicated by vitreous loss. Even relatively innocuous small peripheral round holes may on occasion give rise to RD following cataract surgery.
2. **Myopic** patients are more prone to RD. A break in a myopic eye should be taken more seriously than a similar lesion in a non-myopic eye.
3. **One-eyed** patients with breaks should be taken seriously, particularly if the fellow eye has lost vision from RD.
4. **Family history** may occasionally be relevant; any break or predisposing degeneration should be taken seriously if the patient has a positive family history of RD.
5. **Systemic diseases** associated with an increased risk of RD include Marfan syndrome, Stickler syndrome and Ehlers–Danlos syndrome. Because RD in these patients has a relatively poor prognosis, any break or predisposing degeneration should be treated prophylactically.

Clinical examples (Fig. 12.67a–h)

a. A large equatorial U-tear associated with subclinical RD located in the upper temporal quadrant should be treated prophylactically without delay because the risk of progression to a clinical RD is high. As the tear is located in the upper temporal quadrant, early macular involvement by SRF is likely.

b. A large U-tear in the upper temporal quadrant in an eye with symptomatic, acute PVD should also be treated without delay because the risk of progression to clinical RD is high.

c. An operculated tear bridged by a patent blood vessel should be treated because persistent dynamic vitreoretinal traction on the bridging blood vessel may cause recurrent vitreous haemorrhage.

d. A tear with a free-floating operculum in the lower temporal quadrant detected by chance is much safer because there is no vitreoretinal traction. Prophylaxis is therefore not required in the absence of other risk factors.

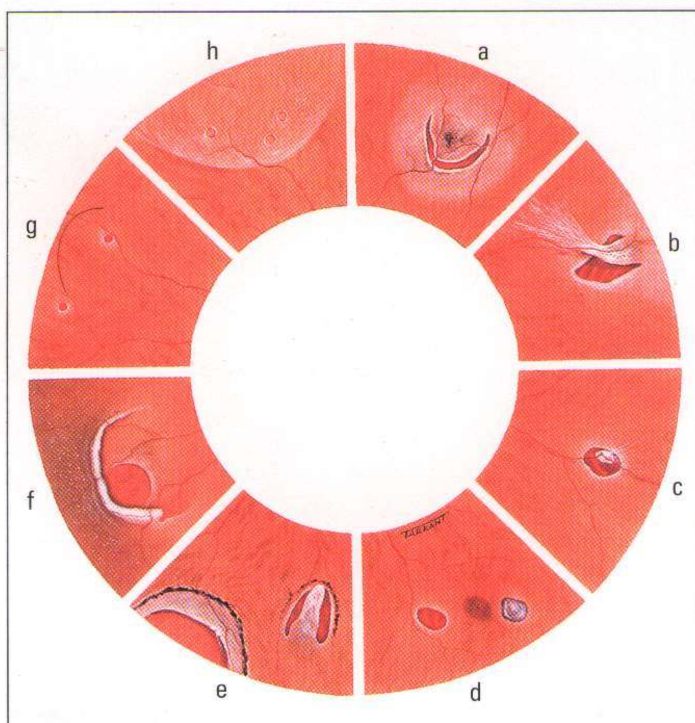


Fig. 12.67
Prophylactic treatment of retinal breaks (*see text*)

e. An inferior U-tear and a dialysis surrounded by pigment detected by chance are both long-standing very low-risk lesions.
f. Degenerative retinoschisis, even with breaks in both layers, does not require treatment. Although this lesion represents a full-thickness defect in the sensory retina, the fluid within the schisis cavity is usually viscid and rarely passes into the subretinal space.
g. Two small asymptomatic holes near the ora serrata do not require treatment; the risk of RD is extremely small as they are probably located within the vitreous base. About 5% of the general population have such lesions.
h. Small inner layer holes in retinoschisis also carry an extremely low risk of RD as there is no communication between the vitreous cavity and the subretinal space.

Predisposing peripheral retinal degenerations

In the absence of associated retinal breaks neither lattice nor snailtrack degenerations merit prophylactic treatment unless associated with one or more of the following risk factors:

1. **RD in the fellow eye** is the most frequent indication.
2. **Aphakia or pseudophakia**, particularly if laser posterior capsulotomy is necessary.
3. **High myopia**, particularly if associated with extensive lattice degeneration.
4. **Strong family history of RD**.
5. **Systemic disease** known to predispose to RD, such as Marfan syndrome, Stickler syndrome and Ehlers–Danlos syndrome.

Treatment modalities

Choice of modality

Prophylactic treatment modalities are (a) *cryotherapy*, (b) *slit-lamp laser photocoagulation* and (c) *indirect ophthalmoscopic laser photocoagulation* combined with scleral indentation. In most cases the choice is based on personal preference and experience as well as the availability of instrumentation. Other considerations include:

1. **Location of lesion**
   - An equatorial lesion can be treated by either photocoagulation or cryotherapy.
   - A post-equatorial lesion can be treated only by photocoagulation unless the conjunctiva is incised.
   - Lesions near the ora serrata can be treated either by cryotherapy or laser photocoagulation using the indirect ophthalmoscope delivery system combined with indentation. Laser photocoagulation using a slit-lamp delivery system is difficult in these cases and it may be impossible to adequately treat the base of a U-tear.
2. **Clarity of media**. Eyes with hazy media are much easier to treat by cryotherapy.
3. **Pupil size**. Eyes with small pupils are easier to treat by cryotherapy.

Slit-lamp laser photocoagulation

1. **Technique**
   a. Laser settings are 200 μm spot size and duration of 0.1–0.2 seconds.
   b. A triple-mirror or a wide field (panfundoscopic) lens is inserted under topical anaesthesia.
   c. The lesion is surrounded by two rows of confluent burns of moderate intensity (Figs 12.68, 12.69).
   d. After treatment the patient is advised to avoid strenuous physical exertion for about 7 days until an adequate adhesion has formed and the lesion is securely sealed.

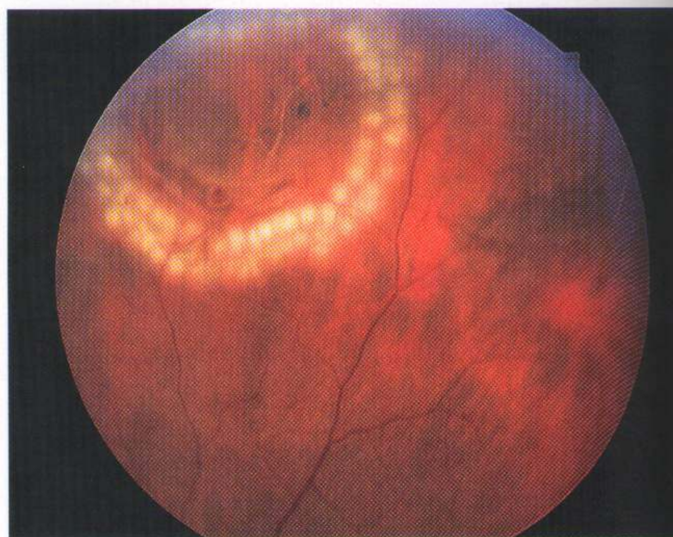


Fig. 12.68
Appearance soon after prophylactic laser photocoagulation of a retinal break

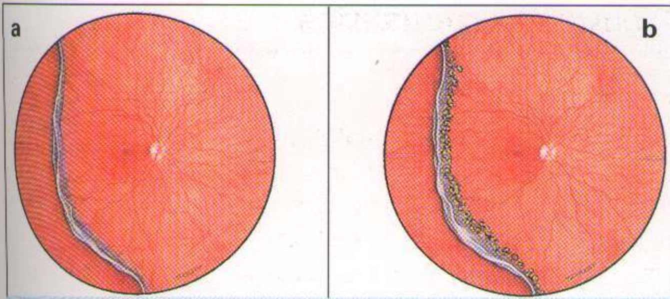


Fig. 12.69
(**a**) Large traumatic retinal dialysis; (**b**) several weeks after prophylactic laser photocoagulation

2. **Potential problems**. Serious complications from peripheral retinal photocoagulation are uncommon and usually associated with excessively heavy treatment to large areas of the retina.
   - Maculopathy in the form of cystoid macular oedema or macular pucker (*see* Fig. 12.101).
   - Choroidal detachment, which may be complicated by secondary angle-closure glaucoma as a result of a forward rotation of the ciliary body.
   - Exudative RD, which usually resolves within 1 or 2 weeks.
   - Rhegmatogenous RD caused by secondary tear formation is very rare.
   - Retinal haemorrhage is rare and can usually be stopped by pressing the contact lens against the eye in order to increase intraocular pressure.

Cryotherapy

1. **Technique**
   a. The eye is anaesthetized with a pledget soaked in amethocaine or a subconjunctival injection of lignocaine in the quadrant of the lesion.
   b. For post-equatorial lesions, a small conjunctival incision may be necessary to enable the cryoprobe to reach the required location.
   c. While viewing with the indirect ophthalmoscope, the sclera is gently indented with the tip of the probe.
   d. The lesion is surrounded with a single row of cryo-applications; freezing is terminated as soon as the retina whitens.
   e. The cryoprobe should not be removed until it has defrosted completely because premature removal may 'crack' the choroid and give rise to a choroidal haemorrhage.
   f. The eye is padded for about 4 hours to prevent chemosis and the patient advised to refrain from strenuous physical activity for a week.

   For about 2 days the treated area appears whitish due to oedema. After about 5 days pigmentation begins to appear. Initially the pigment is fine; later it becomes coarser and is associated with a variable amount of chorioretinal atrophy (Fig. 12.70).
2. **Potential problems**
   - Chemosis and lid oedema are common and innocuous.
   - Transient diplopia may occur as a result of freezing of an extraocular muscle.

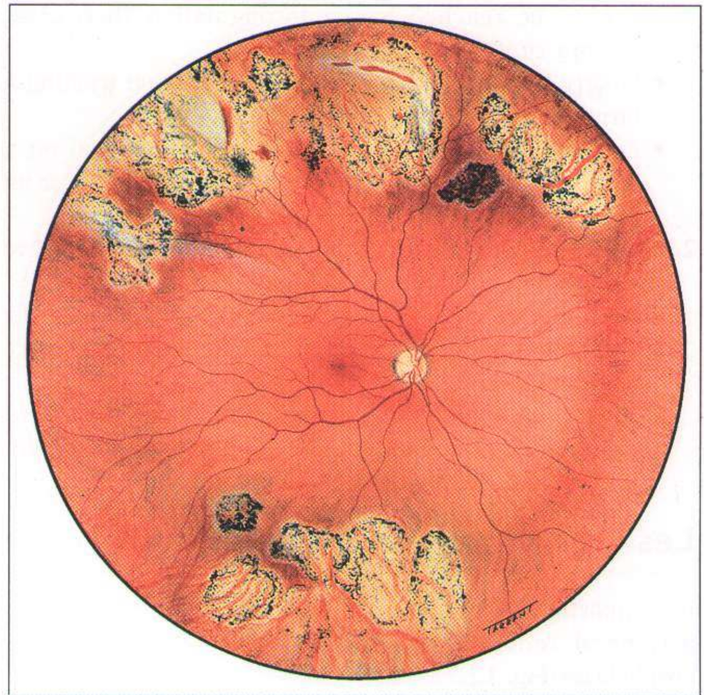


Fig. 12.70
Pigmentation and chorioretinal atrophy following prophylactic cryotherapy

   - Vitritis may occur as a result of excessively heavy treatment.
   - Maculopathy is rare.

Causes of failure

The two main causes of failure of prophylaxis are (a) *inadequate treatment* and (b) *new break formation.*

1. **Inadequate treatment** may be due to the following:
   - Failure to surround the lesion with two rows of burns, particularly the base of a U-tear, is the most common cause of failure. If the most peripheral part of the tear

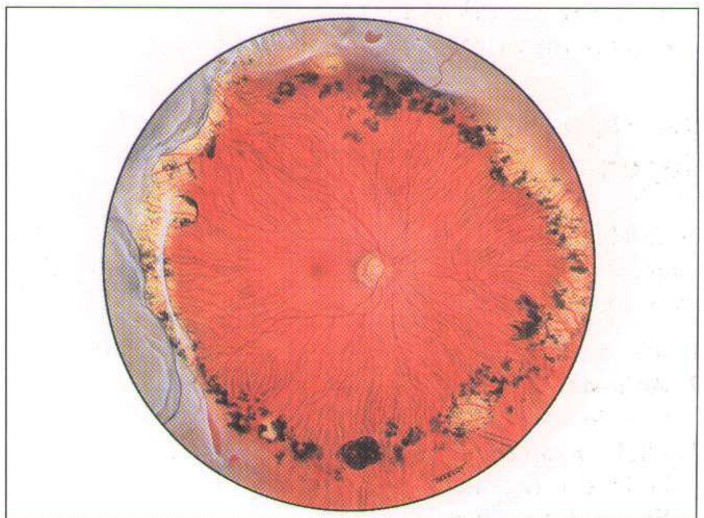


Fig. 12.71
New breaks at 7 and 12 o'clock with subretinal fluid following extensive cryotherapy of lattice degeneration

cannot be reached by photocoagulation then cryotherapy should be employed.
- Failure to apply contiguous treatment when treating a large break or a dialysis.
- Failure to release dynamic vitreoretinal traction on a large U-tear by inserting an explant and failure to use an explant in an eye with a subclinical RD (*see* below).

2. **New break formation** may occur in the following two sites:
   - Within or adjacent to the treated area, usually caused by excessively heavy treatment, particularly of lattice degeneration (Fig. 12.71).
   - In 'normal' appearing retina despite adequate treatment of the predisposing lesion. This is one of the limitations of prophylactic treatment.

Lesions not requiring prophylaxis

It is important to recognize the following entirely innocuous peripheral retinal degenerations which do not require prophylaxis (Fig. 12.72a–f):

a. **Microcystoid degeneration** consists of tiny vesicles with indistinct boundaries on a greyish-white background which make the retina appear thickened and less transparent (*see* Fig. 12.5).

b. **Snowflakes** are minute glistening yellow-white dots which are frequently found scattered diffusely in the peripheral fundus. Foci composed solely of snowflakes are innocuous and require no treatment.

> *NB:* Snowflakes are, however, of considerable clinical importance because, as mentioned previously, they are frequently associated with lattice degeneration, snailtrack degeneration and acquired retinoschisis.

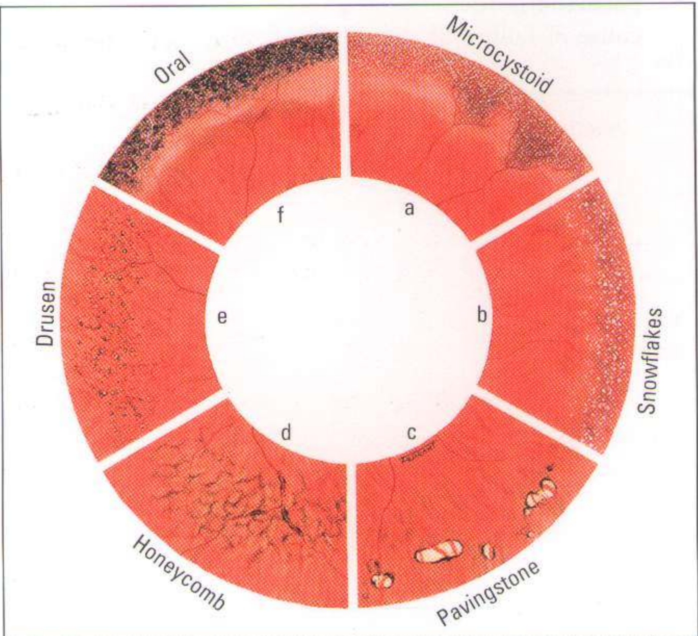


Fig. 12.72
Benign peripheral retinal degenerations

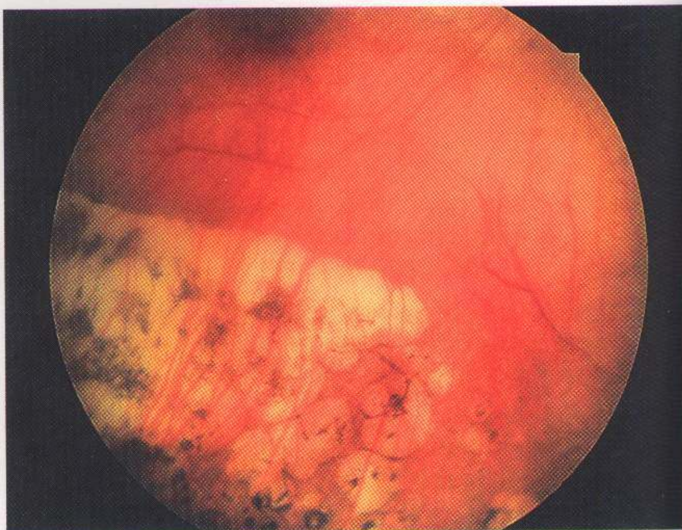


Fig. 12.73
Pavingstone degeneration

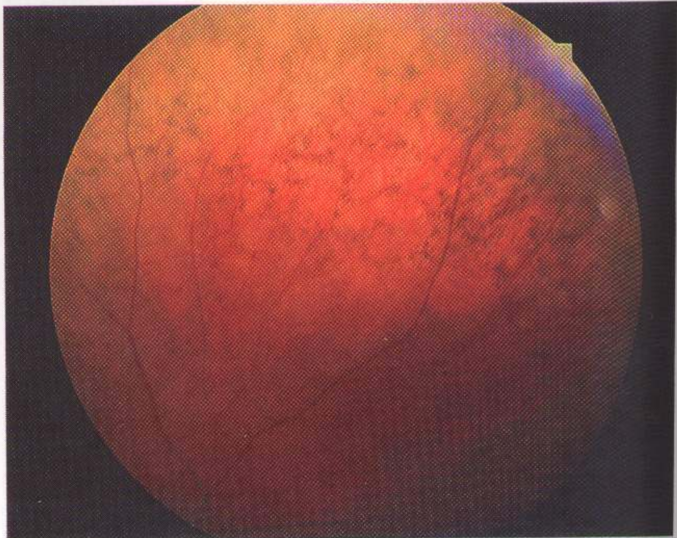


Fig. 12.74
Honeycomb degeneration

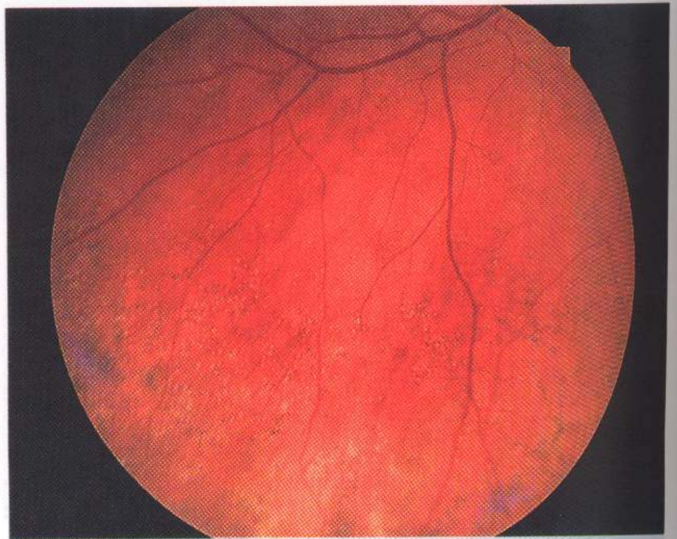


Fig. 12.75
Peripheral drusen

c. **Pavingstone degeneration** is characterized by discrete yellow-white patches of focal chorioretinal atrophy, present to some extent in 25% of normal eyes (Fig. 12.73).
d. **Honeycomb (reticular) degeneration** is an age-related change characterized by a fine network of perivascular pigmentation which may extend posterior to the equator (Fig. 12.74).
e. **Drusen** (colloid bodies) are clusters of small pale lesions which may have hyperpigmented borders (Fig. 12.75).
f. **Oral pigmentary degeneration** is an age-related change consisting of a hyperpigmented band running adjacent to the ora serrata.

Standard retinal surgery

Prognosis for central vision

The main factor governing eventual visual function, provided the retina has been successfully reattached, is the duration of macular involvement by the RD.

- Most eyes with macula-on RD maintain their preoperative visual acuity.
- Within the first week after development of macula-off RD, delay in surgery does not adversely affect visual outcome.
- Macula-off RD of less than 2 months duration results in some impairment of visual acuity but there is no direct correlation between the duration of macular detachment and final visual acuity.
- Macula-off RD of over 2 months duration results in poor visual acuity which appears to be related to the duration of macular involvement.

Principles of scleral buckling

Scleral buckling involves the creation of inward indentation of the sclera ('buckle'). The two main purposes are: (a) *to close retinal breaks* by apposing the RPE to the sensory retina and (b) *to reduce dynamic vitreoretinal traction* at sites of local vitreoretinal adhesion. An explant is material sutured directly onto the sclera to create a buckle.

Local explants

1. **Configuration**
   a. *Radial* explants are placed at right angles to the limbus (Fig. 12.76a).
   b. *Circumferential* explants are placed parallel to the limbus to create a segmental buckle (Fig. 12.76b).
2. **Dimensions**. In order to adequately seal a retinal break it is essential that the buckle be positioned accurately and is of adequate length, width and height.
   a. ***The width*** of a *radial buckle* depends on the width (distance between anterior horns) of the retinal tear and its length depends on the length (distance between base and apex) of the tear. In general, the dimensions of the explant should be twice that of the tear. The required width and length of a *segmental circumferential buckle* depend on the length and width, respectively, of the tear.
   b. ***The height*** is determined by the following interrelated factors:
      - The greater the diameter of the explant, the higher the buckle.
      - The greater the separation of sutures, the higher the buckle.
      - The tighter the sutures over the explant, the higher the buckle.
      - The lower the intraocular pressure, the higher the buckle.
3. **Indications for radial buckling**
   - Large U-tears because there is less tendency to 'fishmouth' (*see* below).
   - Relatively posterior breaks because sutures are easier to insert.
4. **Indications for segmental circumferential buckling**
   - Multiple breaks located in one or two quadrants.
   - Anterior breaks because they can be closed more easily.
   - Wide breaks such as dialyses.

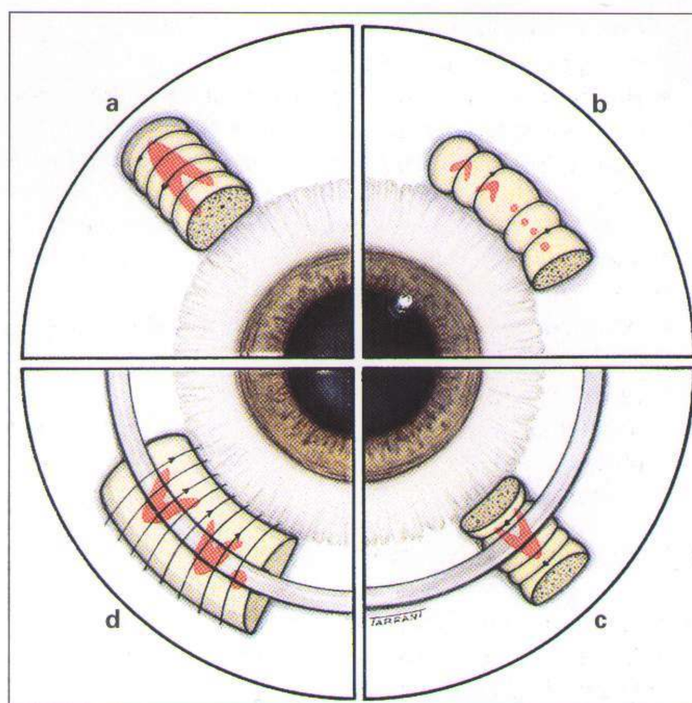


Fig. 12.76
Configuration of scleral buckles. (**a**) Radial sponge; (**b**) circumferential sponge; (**c**) encirclement augmented by a radial sponge; (**d**) encirclement augmented by a solid silicone tyre

Encircling explants

Encircling explants are placed around the entire circumference of the globe to create a 360° buckle (Fig. 12.76c and d).

1. **Dimensions**. Straps with a diameter of 2 mm (no. 40) are most frequently used. A strap induces a fairly narrow buckle, and therefore often has to be supplemented with either radial sponges or circumferential solid silicone tyres to support large tears. A 2 mm high buckle may be produced by tightening the strap by about 12 mm. In contrast to a local explant, the buckle produced by a strap is permanent.
2. **Indications**
   - Breaks involving three or more quadrants.
   - Lattice or snailtrack degeneration involving three or more quadrants.
   - Extensive RD without detectable breaks, particularly in eyes with hazy media.
   - Failed local procedures in which the reason for failure is not apparent.

Drainage of subretinal fluid

Drainage of SRF affords immediate apposition between the sensory retina and RPE. Although a large proportion of RDs can be treated successfully with non-drainage techniques, drainage may be required under certain circumstances. This measure is however, not without potential complications (*see* below). Although non-drainage avoids most of these complications it often does not accomplish immediate apposition between the sensory retina and RPE with flattening of the fovea. If such apposition is delayed for more than 5 days, a satisfactory adhesion will not develop around the retinal break because the 'stickiness' of the RPE will have worn off. This may result in non-attachment of the retina or, in some cases, reopening of the break during the postoperative period. In addition, drainage of SRF allows the use of a large bubble of an internal tamponading agent (air or gas).

Scleral buckling techniques

Preliminary steps

1. With spring-scissors, conjunctiva and Tenon capsule are undermined and cut circumferentially at the limbus in the quadrant(s) corresponding to the retinal breaks.
2. A squint hook is inserted under the corresponding rectus muscles (Fig. 12.77) and bridle sutures are placed.
3. The sclera is inspected to detect thinning (Fig. 12.78) or anomalous vortex veins which may influence subsequent suture placement and drainage of SRF.
4. A 5/0 Dacron partial-thickness scleral suture is inserted at the site calculated to correspond to the apex of the tear.
5. The cut suture is grasped with curved mosquito forceps as close to the knot as possible (Fig. 12.79).
6. While viewing with the indirect ophthalmoscope, the sclera is indented by rotating the forceps. If the indentation does not coincide with the break, the procedure is repeated until accurate localization is achieved.
7. The sclera is gently indented with the tip of the cryoprobe (Fig. 12.80) and freezing is activated until the break is surrounded by a 2 mm margin of whitening.

Insertion of local explant

1. An appropriate-sized explant is selected according to the aforementioned criteria.
2. With calipers, the required distance separating the suture bites is measured and marked on the sclera with cautery.

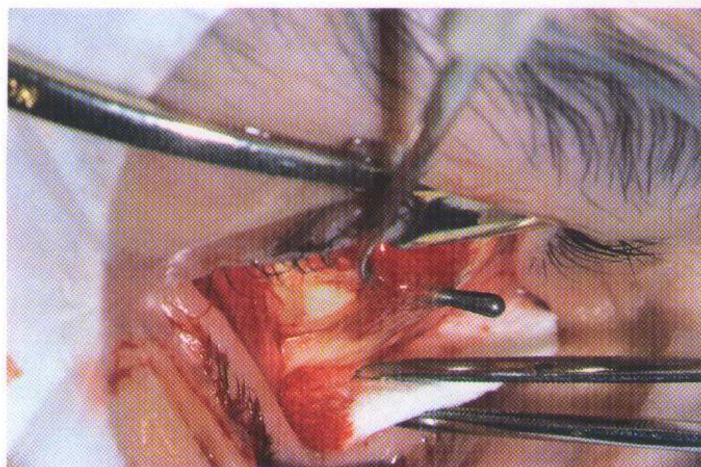


Fig. 12.77
Insertion of squint hook under a rectus muscle

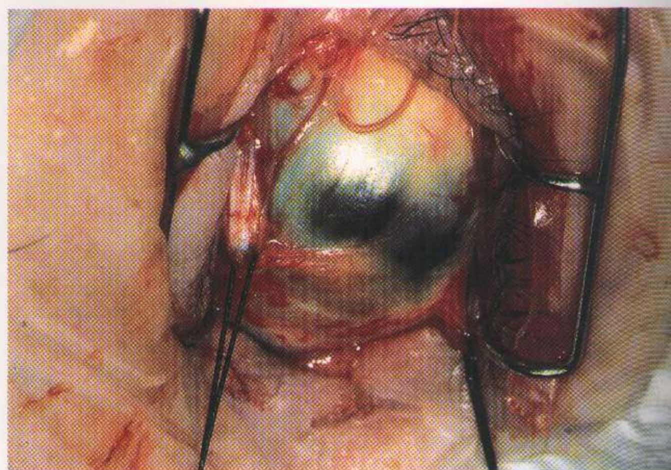


Fig. 12.78
Severe scleral thinning

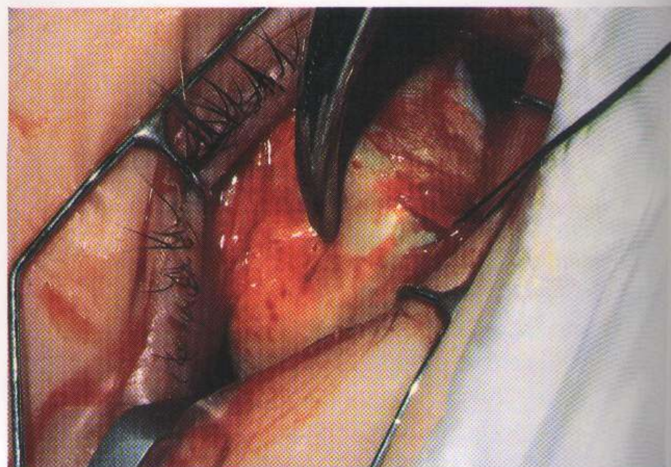


Fig. 12.79
Localizing suture grasped with mosquito forceps

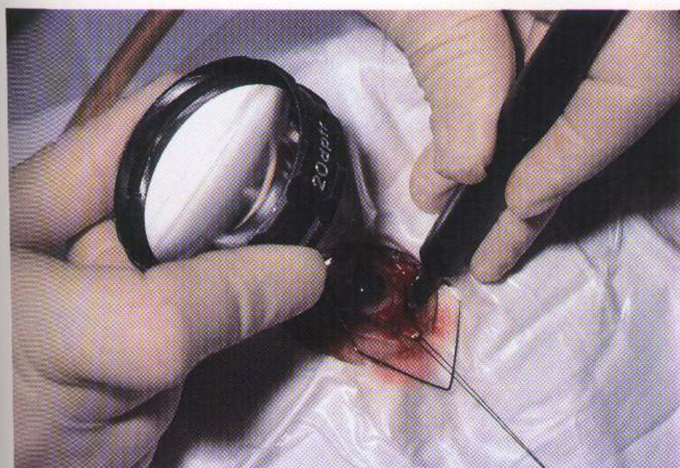


Fig. 12.80
Cryotherapy

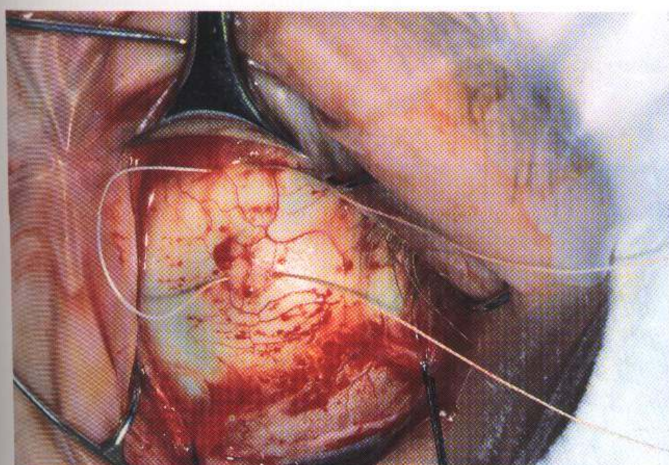


Fig. 12.81
Mattress suture in place



Fig. 12.82
Indentation in relation to the tear—in this case the indentation is too anterior

NB: As a general rule, the separation of sutures should be about one and a half times the diameter of the explant.

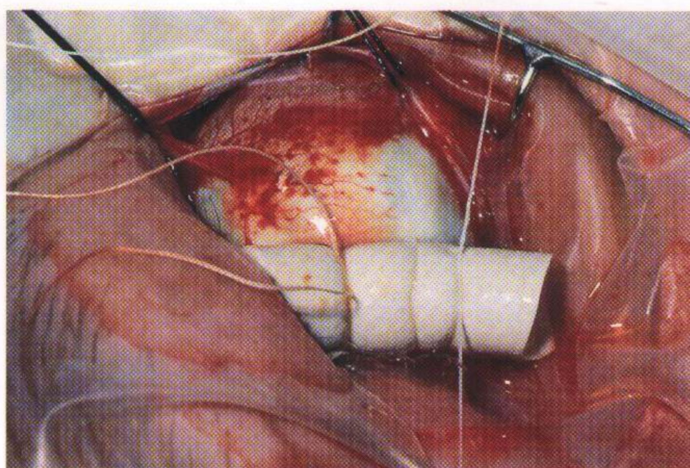


Fig. 12.83
Tying of sutures over sponge

3. A mattress-type suture is inserted which will straddle the explant (Fig. 12.81).
4. SRF is drained, if appropriate (*see* below).
5. The position of the break in relation to the buckle is checked (Fig. 12.82) and the buckle is repositioned if necessary.
6. The sutures are tightened over the explant (Fig. 12.83).

Drain-Air-Cryo-Explant (D-A-C-E)

Localization of relatively anterior breaks in eyes with shallow SRF is easy. However, accurate localization may be difficult or impossible in eyes with bullous RDs, especially if the breaks are post-equatorial. The D-A-C-E technique is useful under these circumstances:

1. SRF is drained to bring retina (and thus the break) closer to the RPE.
2. Air is injected into the vitreous cavity to counteract the hypotony induced by drainage.
3. Breaks may then be accurately localized and treated with cryotherapy.
4. The explant is inserted.

Encircling procedure

1. A strap of appropriate diameter is selected.
2. One end of the strap is grasped with curved mosquito forceps and fed under the four recti (Fig. 12.84).
3. The two ends are secured with a Watzke sleeve in the quadrant of commencement (Fig. 12.85).
4. The strap is tightened by pulling on the two ends (Fig. 12.86) until it fits snugly around the ora serrata.
5. The strap is slid posteriorly (about 4 mm) and secured in each quadrant with a holding suture (Fig. 12.87).
6. SRF is drained.
7. The strap is tightened further to produce the required degree of indentation as observed with indirect ophthalmoscopy.

NB: An ideal height is about 2 mm. This is achieved by shortening the circumference of the strap by about 12 mm (Fig. 12.88).

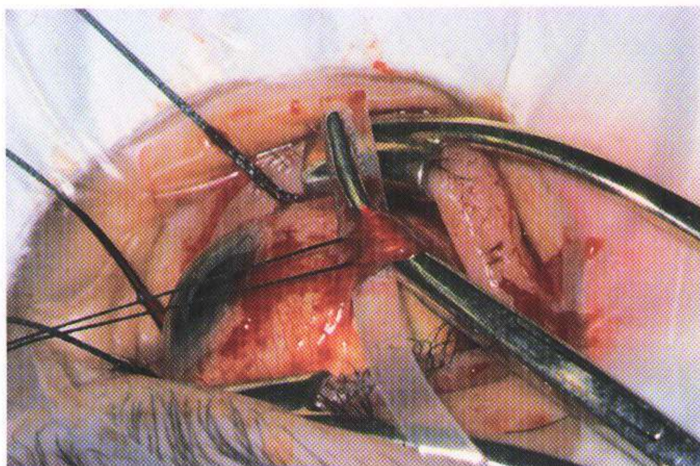


Fig. 12.84
Sliding of end of strap under a rectus muscle



Fig. 12.85
End of strap inserted into Watzke sleeve

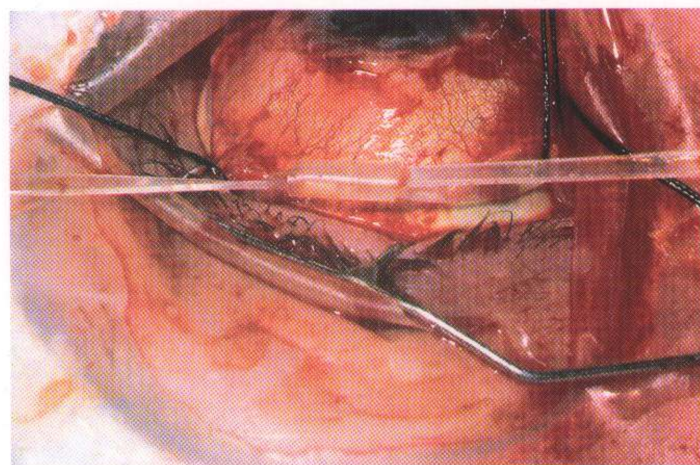


Fig. 12.86
Tightening of strap with Watzke sleeve in position

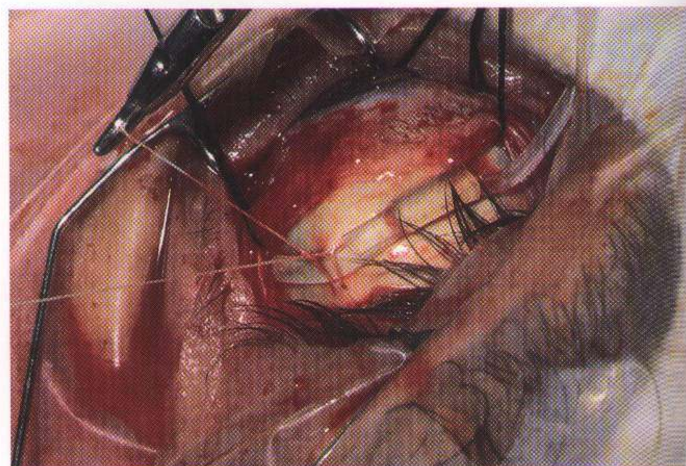


Fig. 12.87
Holding suture in place

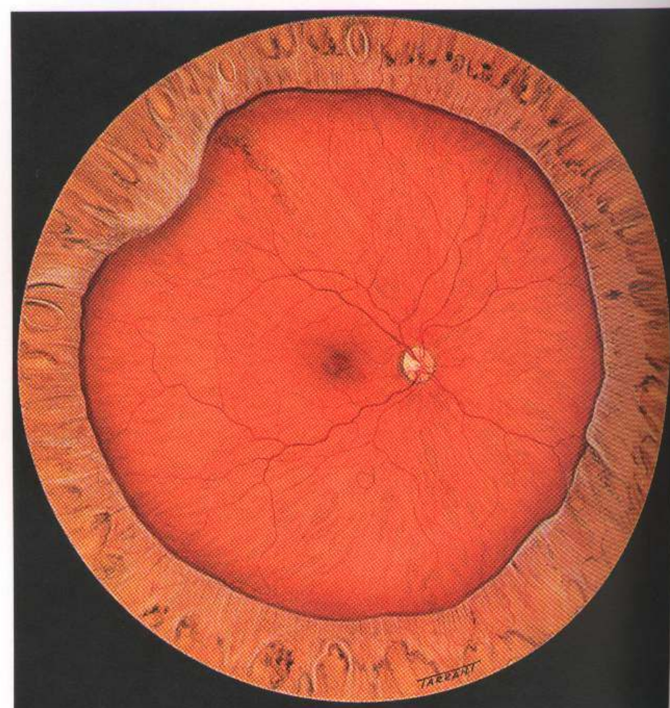


Fig. 12.88
Indentation following encirclement

8. A circumferential buckle should be created such that the retinal breaks 'sit' on the anterior slope of the indent (i.e. the buckle should be placed just behind the breaks).
9. If appropriate, a radial sponge to support a large U-tear (*see* Fig. 12.76c), or a circumferential tyre to support several breaks (*see* Fig. 12.76d) may be inserted under the strap, ensuring that the buckle involves the vitreous base anteriorly.

Drainage of SRF

1. Indications

- Difficulty in localization of breaks in bullous detachments, particularly if the breaks are post-equatorial.
- Immobile retina (e.g. PVR) because a non-drainage procedure will be successful only if the detached retina is sufficiently mobile to move back against the buckle during the postoperative period.
- Long-standing RD because the SRF is viscid and may take months to absorb. Drainage may therefore be necessary, even if the break can be closed without it.
- Inferior RD associated with equatorial tears should ideally be drained because, when the patient assumes the upright position postoperatively, any residual SRF will gravitate inferiorly and may reopen the break.

2. Techniques of drainage have not been standardized. The two most popular methods are described:

a. Method A

- External pressure on the globe is minimized by relaxing traction sutures and lifting the speculum.
- A radial 4 mm sclerotomy is performed, ideally over the area of deepest SRF, and a knuckle of choroid is prolapsed.
- The choroidal knuckle is perforated tangentially with a 25-gauge hypodermic needle on a syringe or a suture needle held by a needle holder (Fig. 12.89).

b. Method B

- The perforation is made directly through sclera, choroid and RPE with the tip of a 27-gauge hypodermic needle bent 2 mm at the tip in a single, swift but controlled fashion.
- In order to prevent haemorrhage from the drainage site, external digital pressure is applied to the globe until the central retinal artery is occluded and complete blanching of the choroidal vascular bed is achieved.
- Pressure is maintained for 5 minutes and the fundus is re-examined; if bleeding is persistent, pressure is reapplied for a further 2 minutes.

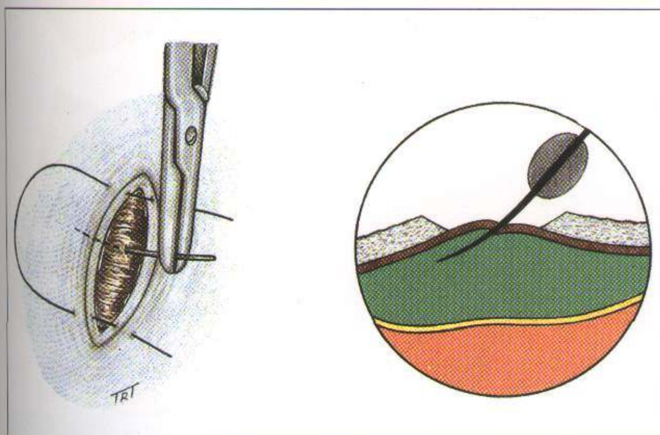


Fig. 12.89
Drainage of subretinal fluid

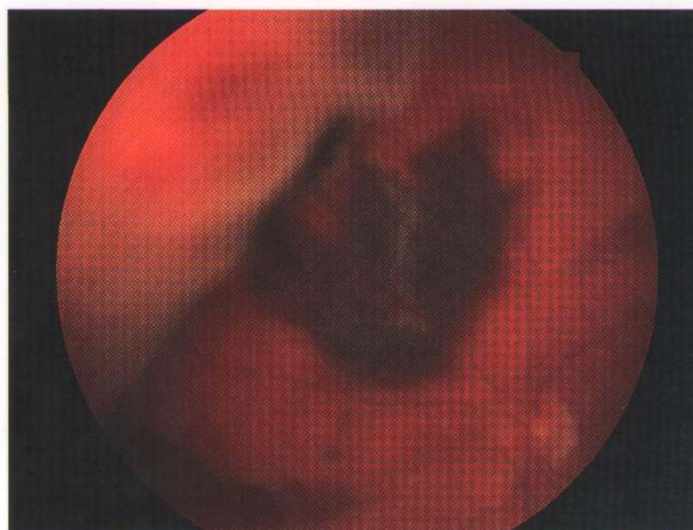


Fig. 12.90
Subretinal haemorrhage associated with drainage of subretinal fluid

3. Complications

- Haemorrhage (Fig. 12.90), usually due to perforation of a large choroidal vessel.
- Dry tap, in which failure of drainage may be due to incarceration of intraocular structures in the ostium.
- Iatrogenic break formation caused by perforating the retina during drainage.
- Retinal incarceration (Fig. 12.91) is a serious problem, which frequently leads to failure to reattach the retina.
- 'Fishmouthing' is a tendency for U-tears to paradoxically open widely following scleral buckling and drainage of SRF. A tear may communicate with a radial retinal fold rendering it difficult to close (Fig. 12.92a). Management of this problem involves the insertion of

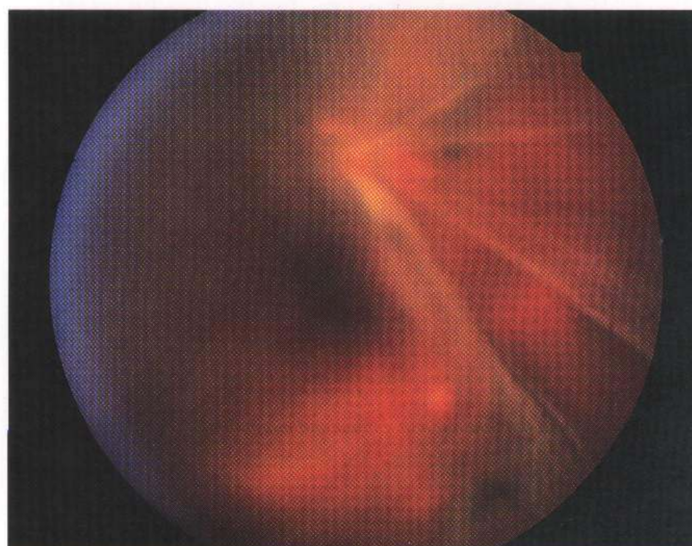


Fig. 12.91
Retinal incarceration into the drainage site

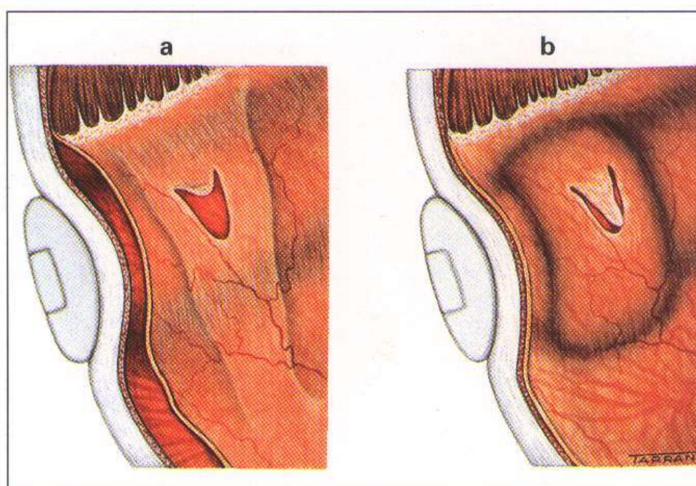


Fig. 12.92
(**a**) 'Fishmouthing' of a U-tear communicating with a radial fold; (**b**) insertion of radial buckle

an additional radial buckle and injection of air into the vitreous cavity (Fig. 12.92b).

Intravitreal air injection

1. Indications
- Severe ocular hypotony after drainage of SRF.
- 'Fishmouthing' of a U-shaped tear.
- Radial retinal folds.

2. Technique
a. A 25-gauge needle on a 5 ml syringe filled with filtered air is employed.
b. The globe is steadied and the needle inserted 3.5 mm behind the limbus, through the pars plana.
c. While viewing through the pupil using the indirect ophthalmoscope without a condensing lens, the needle is aimed at the centre of the vitreous cavity and pushed forward until it is just visible in the pupil.
d. A single smooth injection is made (Fig. 12.93a).

3. Potential problems
- Loss of visualization of the fundus due to the formation of small air bubbles which may occur if the needle is inserted too deeply into the vitreous cavity (Fig. 12.93b).

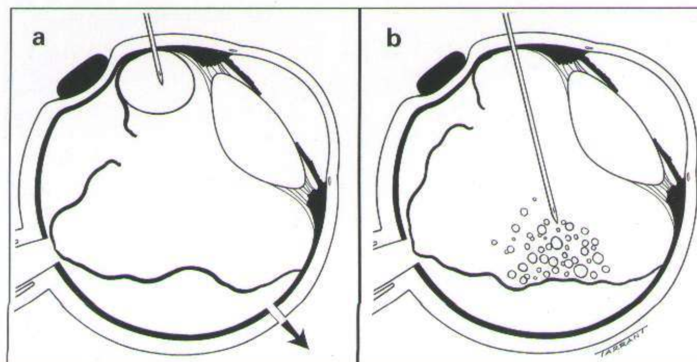


Fig. 12.93
Intravitreal air injection. (**a**) Correct method; (**b**) incorrect method

- Elevation of intraocular pressure by injecting too much.
- Lens damage by the needle, if angled anteriorly.
- Retinal damage may occur if the needle is inserted too posteriorly.

Clinical examples

The following examples emphasize the most important aspects of scleral buckling.

Fresh retinal detachment

1. Preoperative considerations. Examination shows a localized right upper temporal RD due to a U-tear (Fig. 12.94a). The prognosis for central vision is good because the macula is uninvolved. The patient should be admitted immediately, rested supine and operated on as soon as possible because the macula is in great danger for two reasons:
- The break is located in the upper temporal quadrant.
- Subretinal fluid will spread quickly because the break is large.

2. Surgical technique
a. The peritomy should extend from 8.30 to 12.30 o'clock to expose the lateral and superior recti.
b. Most U-tears can be sealed with a 5 mm sponge explant. The suture bites should be about 8 mm apart to obtain adequate height to the buckle. The buckle should be placed radially to prevent the possibility of 'fishmouthing' (Fig. 12.94b). Figure 12.94c illustrates an undersized buckle. Accurate positioning of the explant is vital in this case. Figure 12.94d shows a malpositioned buckle.

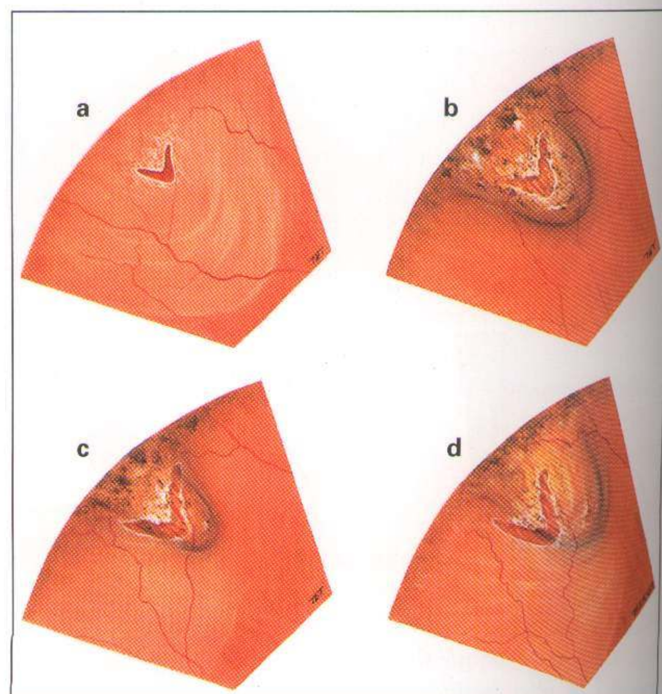


Fig. 12.94
Management of fresh upper temporal retinal detachment and causes of failure (*see text*)

c. Drainage of SRF is not required because:
- The retina is freely mobile.
- The break can be apposed to the RPE without difficulty.
- The SRF is watery because the RD is fresh.

> *NB:* Great care should be taken not to occlude the central retinal artery during a non-drainage procedure.

Long-standing retinal detachment

1. **Preoperative considerations**. Examination shows an extensive right 'macula-off' RD associated with a U-tear in the upper temporal quadrant and two small round holes in the lower temporal quadrant (Fig. 12.95a). A partially pigmented demarcation line is present at the junction of detached and flat retina, and a secondary intraretinal cyst is present inferiorly. This is therefore a long-standing RD because demarcation marks take about 3 months to develop and secondary retinal cysts usually take about 12 months. The prognosis for restoration of good visual acuity is poor because the macula has probably been detached for at least 12 months. There is therefore no urgency for surgery, which can be performed at the patient's and surgeon's convenience.
2. **Surgical technique**
   a. The peritomy should extend from 5.30 to 12.30 o'clock to expose the superior, lateral and inferior recti.
   b. The U-tear can be sealed with a 5 mm wide radial explant and the two holes with a 4 mm wide circumferential segmental explant (Fig. 12.95b). Alternatively, all breaks can be sealed with a long 4 mm wide circumferential segmental sponge explant extending from 7 to 10.30 o'clock (Fig. 12.95c).
   c. Drainage of SRF is required because the SRF is viscid and likely to take a long time to absorb.

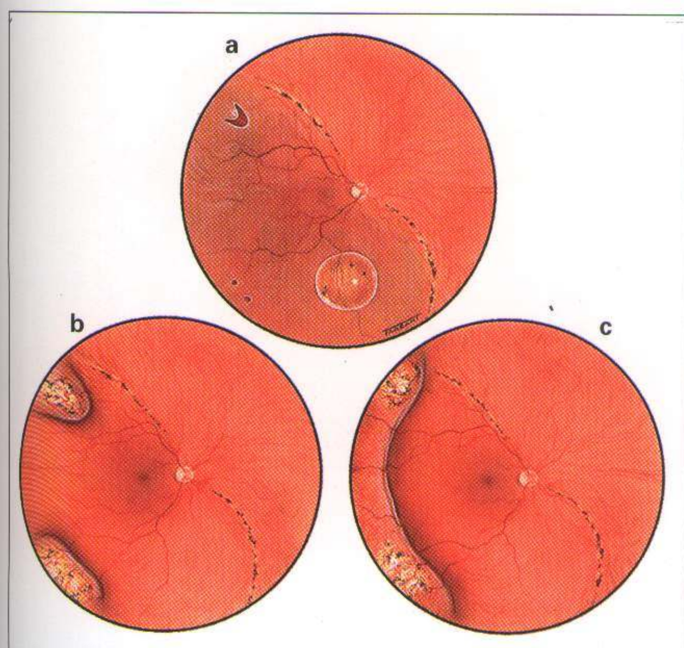


Fig. 12.95
Management of long-standing retinal detachment (*see text*)

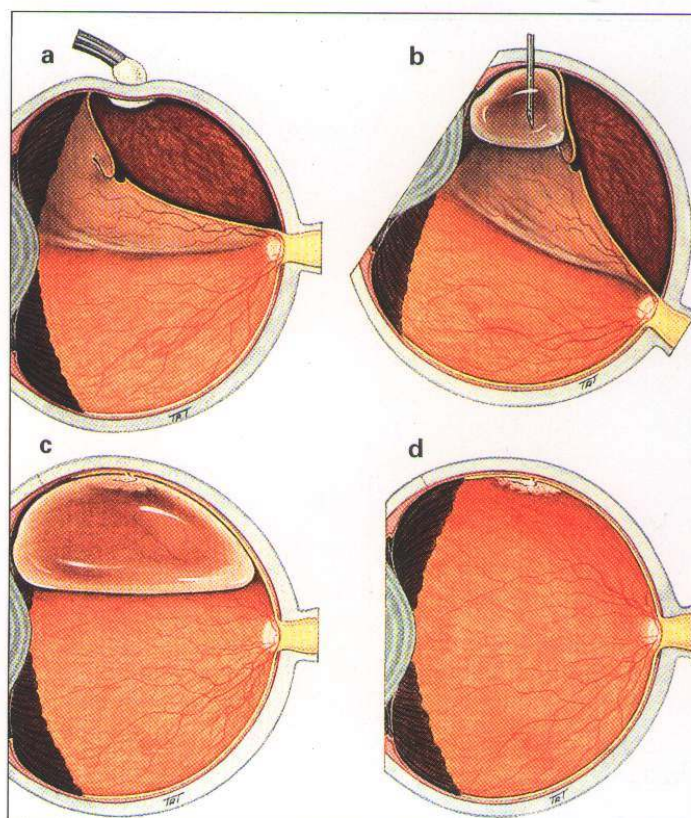


Fig. 12.96
Pneumatic retinopexy (*see text*)

Pneumatic retinopexy

Pneumatic retinopexy is an out-patient procedure in which an intravitreal expanding gas bubble is used to seal a retinal break and reattach the retina without scleral buckling. The most frequently used gases are sulphur hexafluoride (SF_6) and perfluoropropane (C_3F_8).

1. **Indications** are uncomplicated RDs with a small retinal break or a cluster of breaks extending over an area of less than two clock hours situated in the upper two-thirds of the peripheral retina.
2. **Surgical technique**
   a. Retinal breaks are treated with cryotherapy (Fig. 12.96a).
   b. An intravitreal injection is made of either 0.5 ml of 100% SF_6 or 0.3 ml of 100% C_3F_8 (Fig. 12.96b).
   c. Postoperatively, the patient is positioned so that the break is uppermost and the rising gas bubble remains in contact with the tear for 5–7 days (Fig. 12.96c and d).
   d. If required, additional cryotherapy or photocoagulation may be applied around the break.

Causes of failure

Early failure

This is most frequently due to the presence of an open break. The causes can be preoperative or operative.

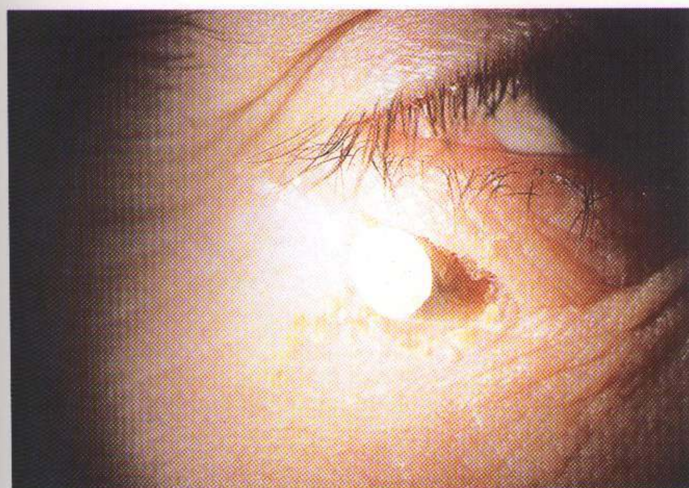


Fig. 12.100
Erosion of sponge through lower lid

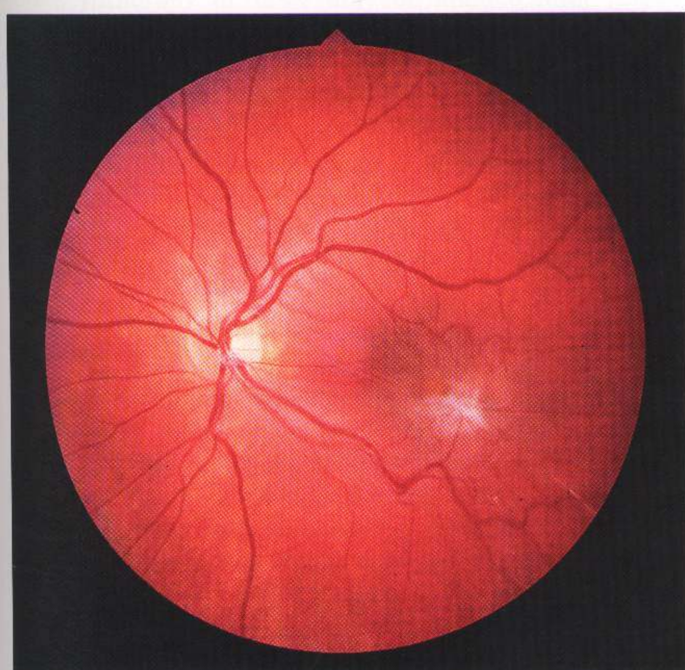


Fig. 12.101
Macular pucker

3. **Erosion** through the skin is very rare (Fig. 12.100).

Maculopathy

1. **Cellophane maculopathy** is characterized by an abnormal reflex at the macula, unassociated with distortion of the surrounding blood vessels. This finding is compatible with normal visual acuity.
2. **Macular pucker** is characterized by an opaque epiretinal membrane with distortion of blood vessels (Fig. 12.101). This complication appears unrelated to the type, extent or duration of RD, or to the type of surgical procedure. Most eyes with macular pucker have a visual acuity of less than 6/18.
3. **Pigmentary maculopathy** is usually the result of excessive cryotherapy.
4. **Atrophic maculopathy** is usually caused by the gravitation of blood in the subretinal space due to operative choroidal haemorrhage. This is seen in association with drainage of SRF since the needle track allows the blood to enter the subretinal space.

Diplopia

Transient diplopia is common during the immediate postoperative period and is a good prognostic sign indicating macular reattachment. Persistent diplopia is rare and may necessitate strabismus surgery or botulinum toxin injection. The following are the main predisposing factors:

- A large explant inserted under a rectus muscle. In most cases the diplopia resolves spontaneously after a few weeks or months and requires no specific therapy apart from reassurance or the temporary use of prisms. Very rarely a sponge may need to be removed.
- Surgical disinsertion of a rectus muscle (usually superior or inferior rectus) in order to place a buckle under it.
- Rupture of the muscle belly as a result of excessive traction on the sutures.
- Severe conjunctival scarring, usually associated with repeated operations, may cause mechanical restriction of eye movements.
- Decompensation of a large heterophoria resulting from poor postoperative visual acuity in the operated eye.

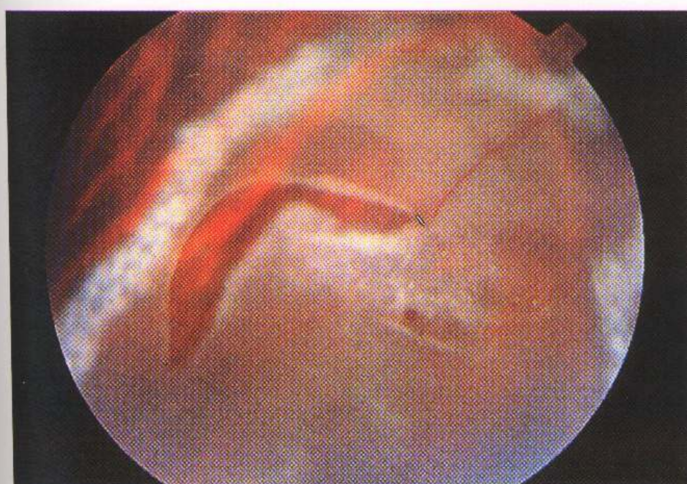
Pars plana vitrectomy

Introduction

Pars plana vitrectomy is a microsurgical procedure designed to remove vitreous gel, usually in order to gain access to a diseased retina. The most common approach is via three separate incisions through the pars plana.

Aims

1. **Excision of the posterior hyaloid face** (PHF) up to the posterior border of the vitreous base is of paramount importance in eyes with RD. The so-called 'core' vitrectomy which leaves the PHF and any associated retinal membranes intact is justifiable only in the management of endophthalmitis.
2. **Relief of vitreoretinal traction** by epiretinal membrane dissection and/or retinotomy.
3. **Retinal manipulation and reattachment**.
4. **Creation of a space** within the vitreous cavity for subsequent internal tamponade.
5. **Miscellaneous** aims, where appropriate, include removal of associated vitreous opacities, cataract, dislocated lens fragments or intraocular foreign bodies.


Fig. 12.105
Posterior retinal tear

- Cryotherapy or laser may be applied after retinal reattachment so that the amount of destructive energy is minimized.
- A tamponading agent ensures postoperative internal closure of retinal breaks.

2. **Complicated RDs** in which retinal breaks cannot be closed by conventional scleral buckling because of large size (Fig. 12.104), posterior location (Fig. 12.105) and association with PVR.

Tractional RD

1. **In proliferative diabetic retinopathy**, vitrectomy is indicated when RD involves or threatens the macula (*see* Chapter 14) and may be combined with internal panretinal laser photocoagulation. Combined tractional–rhegmatogenous RD should be treated urgently, even if the macula is not involved, because SRF is likely to spread quickly to involve the macula.
2. **In penetrating trauma**, vitrectomy is aimed at visual rehabilitation and minimizing the tractional process predisposing to RD.

Techniques

Proliferative vitreoretinopathy

The aims of surgery include release of transvitreal traction by vitrectomy and tangential (surface) traction by membrane dissection in order to restore retinal mobility and allow closure of retinal breaks.

1. **Initial steps**
   a. The infusion cannula is secured to an inferotemporal sclerotomy 3.5 mm behind the limbus.
   b. Two further sclerotomies are made at the 10 and 2 o'clock positions, through which the cutter and the fibre-optic light pipe are inserted (Fig. 12.106).
   c. The central vitreous gel and posterior hyaloid face are excised.

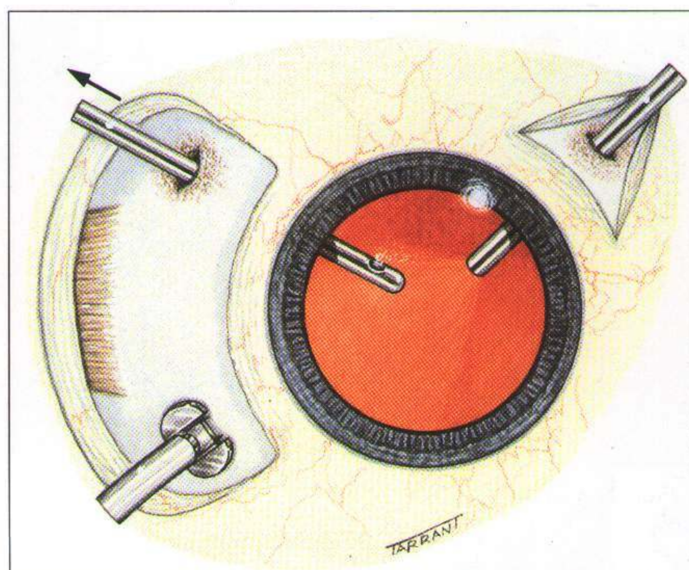


Fig. 12.106
Infusion cannula, light pipe and vitreous cutter in position

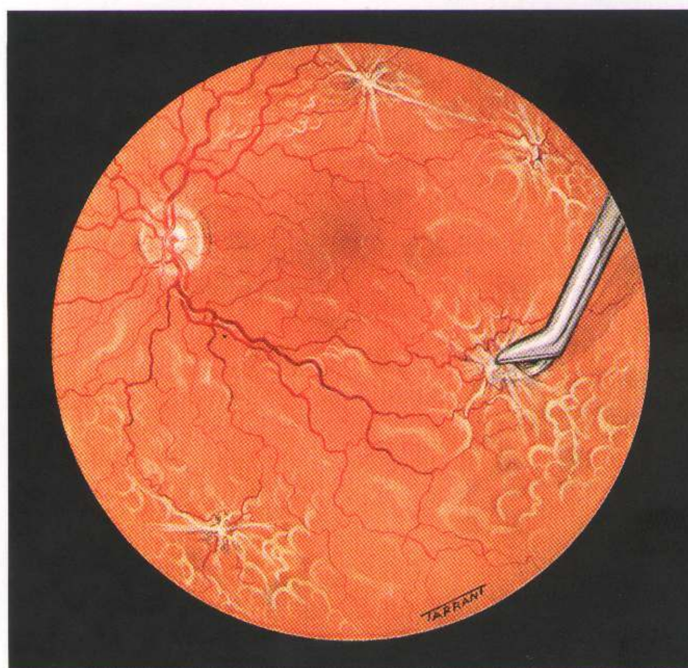


Fig. 12.107
Dissection of starfolds in proliferative vitreoretinopathy

2. **Membrane dissection** of localized fixed retinal (star) folds is as follows:
   a. The tip of a vertically cutting scissors is engaged in the membrane at the edge of the valley between two adjacent retinal folds (Fig. 12.107) and the membrane is pulled towards the ora serrata until it peels from the surface of the retina.
   b. Internal fluid–air exchange is performed and retinopexy applied to retinal breaks.
   c. The vitreous base is supported by a broad scleral buckle.
   d. The intraocular air is exchanged for an extended intraocular tamponading agent such as C_3F_8 or silicone oil.

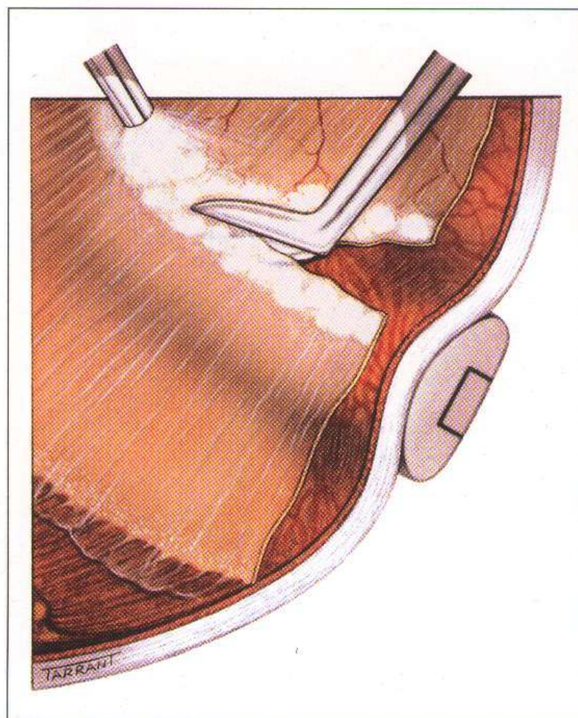


Fig. 12.108
Relieving retinotomy in proliferative vitreoretinopathy

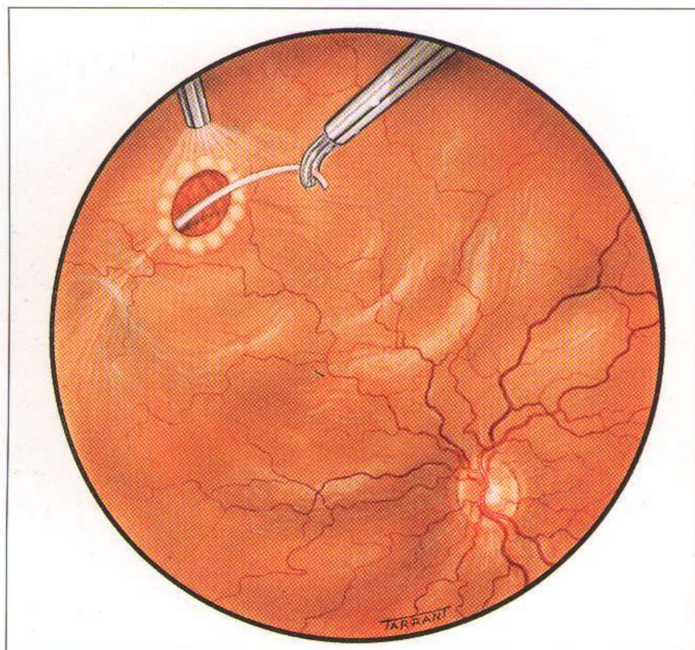


Fig. 12.109
Removal of a subretinal membrane in proliferative vitreoretinopathy

3. **Relieving retinotomy** may be required after membrane dissection if retinal mobility is deemed insufficient for lasting reattachment (Fig. 12.108).
4. **Removal of subretinal membranes** may be required in selected cases (Fig. 12.109).

Tractional retinal detachment

The aims of surgery are to release anteroposterior and/or circumferential vitreoretinal traction. Since the membranes are vascularized they cannot be simply peeled from the surface of the retina as in PVR because this would result in haemorrhage and tearing of the retina. Fibrovascular membranes in diabetic tractional RDs may be removed by:

1. **Delamination**, which involves the horizontal cutting of the individual vascular pegs connecting the membranes to the surface of the retina (Fig. 12.110). This is preferable to segmentation because it allows the complete removal of fibrovascular tissue from the retinal surface (en-bloc delamination).
2. **Segmentation** involves the vertical cutting of epiretinal membranes into small segments (Fig. 12.111). It is used to release circumferential vitreoretinal traction when delamination is difficult or impossible, such as in very mobile combined tractional–rhegmatogenous RD associated with posterior retinal breaks.

Postoperative complications

1. **Raised intraocular pressure** may be caused by the following mechanisms:
   - Overexpansion of intraocular gas.

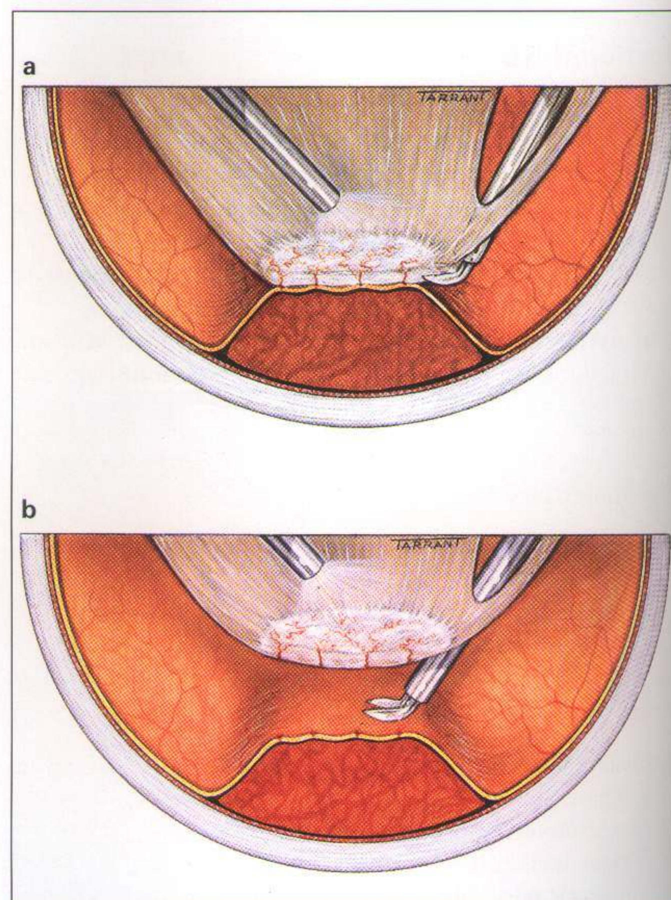


Fig.12.110
(a) Delamination with horizontally cutting scissors; (b) delamination completed

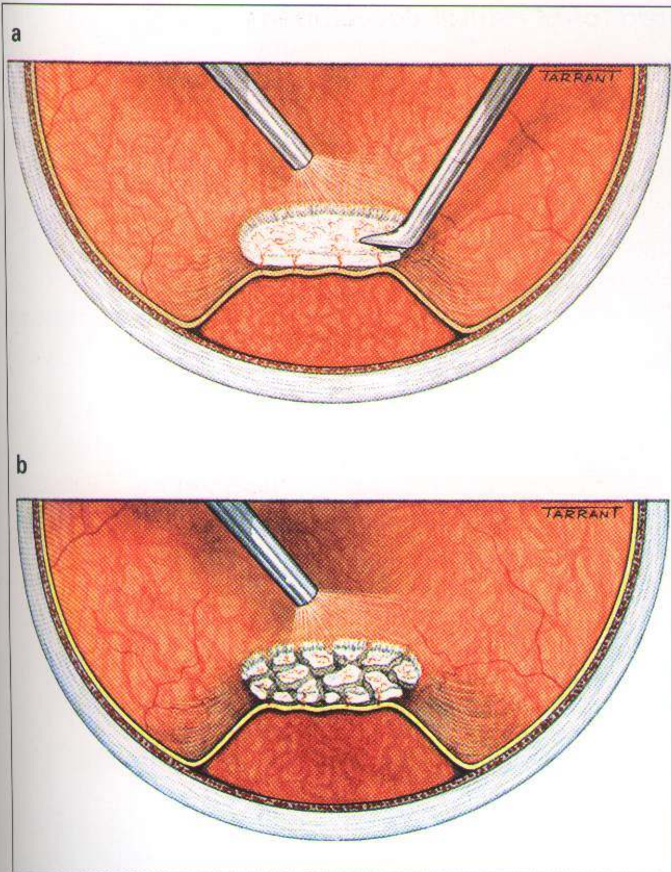


Fig. 12.111
(a) Segmentation with vertically cutting scissors; (b) segmentation completed

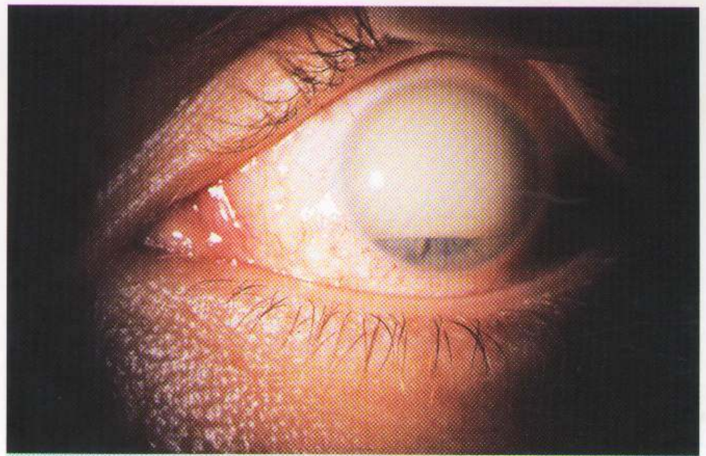


Fig. 12.113
Emulsified silicone oil in the anterior chamber

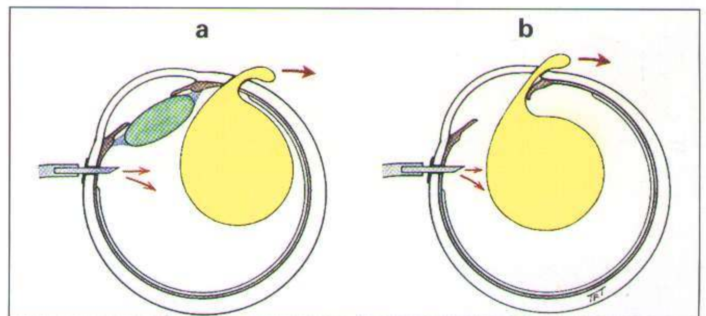


Fig. 12.114
Removal of silicone oil. (**a**) In phakic eye; (**b**) in aphakic eye



Fig. 12.112
Silicone oil in the anterior chamber



Fig. 12.115
Induced cataract and emulsified silicone oil

- Early silicone-oil-induced glaucoma is caused by oil in the anterior chamber (Fig. 12.112).
- Late silicone-oil-induced glaucoma is probably due to trabecular blockage by emulsified oil in the anterior chamber (Fig. 12.113). This may be prevented by early removal of silicone oil either via the pars plana in the phakic eye or the limbus in the aphakic eye (Fig. 12.114).
- Ghost cell or steroid-induced glaucoma.

2. **Cataract** may be caused by the following:
   - Gas-induced lens opacities are usually transient and can be minimized by using lower concentrations and smaller volumes of gas.
   - Silicone-oil-induced lens opacities (Fig. 12.115) develop in almost all phakic eyes. If a cataract develops, the silicone oil may be removed in conjunction with cataract surgery.

- Delayed nuclear sclerosis almost invariably develops within 5–10 years.

3. **Retinal re-detachment** occurs most commonly when the intraocular gas bubble has absorbed (3–6 weeks postoperatively), or following removal of silicone oil. The main causes are:
   - Reopening of the original break because of inadequate operative dissection in eyes with PVR or re-proliferation of epiretinal membranes, more common in eyes with proliferative diabetic retinopathy.
   - New or missed breaks, especially those related to the pars plana sclerotomy sites.

NB: Early removal of silicone oil is associated with a 25% risk of retinal re-detachment in eyes with PVR and giant tears, and 11% risk in eyes with proliferative diabetic retinopathy